THE NEW BORN

JOURNAL OF NNF KERALA

Theme: Neonatal Emergencies

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From the Editorial Desk

Dear Colleagues in Neonatology,

It is with trepidation more than anything else thing else that I pen these lines. Trepidation because,I realise there is a whole lot to improve in this particular 'Neonate' The bar set by my predecessor was so high that I decided the only way forward is to not even try! There was a heartwarming level cooperation from all colleagues as is evident in the choice of invited articles We planned this edition in 3 sections, a Ready-Reckoner section where we have tried to touch upon most key topics; a Mid Neocon 2017 section with abstracts & a final Section the Miscellanea which is filled with case reports & pictures. I say a big thank you to the editorial team with me but hasten to add that all deficiencies are unintentional & solely mine!

Sincerely,

Preetha

Message

Dear friends

I'm pleased to see Dr Preetha and team coming out with first magazine of NNF Kerala 2017.

NNF Kerala has taken a big leap this year by forming 8 district branches with in a short span of 6 months. Strength of an organisation lies is membership and branches. We had a very good start in February when we started traveling all over kerala in connection with new district branch formation. Response from each & every corner was uniformly. encouraging,so much so that we are actually far ahead of our action plansfor this year!

When we started at the grass root level, we have managed to identify a lot of things which require our urgent attention.The neonatal transport system is in very primitive level in kerala.15 states in India have very good neonatal transport system in place whereas in this vital aspect Kerala is literally floundering. We have very good (albeit few in number) tertiary care neonatal intensive care units in kerala. To reach these centres, a sick neonate has to be transported, that too most often in an ordinary vehicle.We have submitted a proposal to the government in this regard on July 1st at the health minister's office in Thiruvananthapuram. I am very very happy to learn that they have already taken initial steps to implement this project.

In Kerala, there are 464 delivery points in private sector and 72 delivery points in public sector. 28% deliveries take place in public sector and the major share of 72 % occurs in private sector. To reduce our IMR we have to reduce NMR, which as we know is the major contributer of IMR. For this we must develop a good neonatal resuscitation team at all delivery points. We,NNF Kerala can easily train the available work force in each delivery point with the help of our state government. This suggestion alsowas well taken by the government and we hope to start our work soon. We now have to accredit all existing neonatal care unit according to the level of care they are expected to deliver. Each & every baby delivered in Kerala should get uniformly expert care irrespective of where they are born.

We do not have any cardiothoracic surgery unit for neonates and children in northern kerala beyond Ernakulam till Kasarakode. We should have at least one in government sector attached to Calicut Medical College. We hope to start our website and E magazine during midneocon in August 2017. Wealso hope to start work on our data bank shortly.

As I wind down this message,I do so with the realisation that we have much much more to do in the field of neonatal care. Hope God almighty will give our team bith courage and strength to achieve more and more during our tenure. I'm grateful to one and all who contributed to the growth and development of NNF in Kerala.

Thank you and best wishes!

Jai NNF Jai IAP

Dr.Santosh M K
President NNF Kerala

Message

Dear Colleagues,

Greeting from NNF Kerala.

Its my pleasure and privilege to write a forward to the NNF Kerala journal with a theme of Neonatal emergencies. NNF Kerala is going through its Golden period. The new team of office bearers are doing a very dedicated team work with SMART goals and this will take NNF Kerala to greater heights. We need your support to achieve this. Let me take this opportunity to congratulate Dr.Preetha for preparing this journal and team Thrissur for organising Mid Neocon 2017.

Best regards

Dr.AK.Jayachandran
MBBS, DCH, MRCPCH, CCT
Secretary.NNF Kerala

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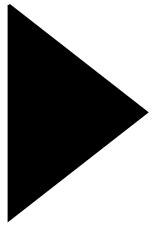


Triage in Emergency Room

Dr Navin Jain

Head Department of Neonatology,
KIMS Hospital, Trivandrum

The doctor or nurse who is the first person seeing the sick baby must first check for life threatening emergencies - apnea, bradycardia, seizure or bleeding that need to be addressed immediately.

Resuscitate first (As in NRP)

- Apnea
- Bradycardia

Treat as emergency

- Seizure
  Airway, **B**reathing, **C**irculation, **D**extrose, and **E**lectrolytes (calcium, sodium)
- Bleeding
  Venous access, assess and treat poor perfusion, arrange blood

If the sick baby doesn't need resuscitation, the doctor or nurse seeing the baby 1st must perform a primary survey before starting therapies, planning investigations or taking history.

Primary survey:

STOPS:

Sensorium and skin color, Temperature, Oxygenation, Perfusion, Sugar is an acronym that reminds the paediatrician important primary assessment parameters. This clinical tool has been in use in the authors NICU and many other Indian NICUs.

The Canadian ACORN also recommends assessment of respiration (O), cardiovascular (P), neurology (S), sugars (S) and need for IVF, temperature regulation (T) and adds need for antibiotics, surgical emergencies and communication with parents in the primary survey.

It takes only 1-2 minutes for a doctor or nurse to answer 5 questions as GOOD or BAD

The primary survey should guide treatment and assessment priorities.

1. A. SENSORIUM

ACTIVE

A neonate who has a good tone, responds briskly to touch, cries normally, moves spontaneously in the observation period would be labelled as active (A).

LETHARGIC

If the neonate is hypotonic, sleeping with poor response to handling, has no spontaneous movements & poor cry, the neonate is labelled as lethargic (L).

1. B. SKIN COLOR

- Pallor / central cyanosis
  urgent need resuscitation
- Peripheral cyanosis - Cold stress
  provide warmth and reassess, BAD if peripheral cyanosis persists after 15 min

2. TEMPERATURE

Peripheries: Use dorsum of your hand to compare temperature of abdomen & hands & feet (both should be warm). Neonates with warm peripheries are unlikely to be seriously sick.

Normal temperature: 36.5 - 37.5 C. Measure axillary temperature. Measurement of rectal temperature not recommended for routine. Measure for 3 minutes (or till beep in digital thermometer); do not add one degree to axillary temperature in neonates.

GOOD

Warm peripheries and normal axillary temperature

BAD

Cool peripheries, that remain cool in spite of covering / rewarming for 15 minutes

Fever (>37.5) / hypothermia (<36.5)

3. OXYGENATION

a. Assess respiratory distress

b. Assess oxygen need

Respiratory distress

Retractions & grunt (severe RD) or fast breathing > 60 / min

Oxygenation

GOOD

The saturation (SpO2) maintained above 90 % without oxygen therapy

BAD

Needs oxygen to maintain saturation (SpO2) above 90 %

Moderately sick: Needs 30 % oxygen (head box with both port holes open)

Critically sick: Needs 60 % oxygen or more (need to close one port hole or both of the head box oxygen)

4. PERFUSION

GOOD

Active baby, warm peripheries, normal capillary refill time (<3 seconds), normal heart rate (120- 140 bpm), well felt pulses

BAD

If a neonate has poor perfusion, the neonate is already seriously sick.

Early signs of poor perfusion (compensated shock)

a. Cool peripheries (hands & feet) felt by dorsum of your hand (despite covering / warmer for 15 minutes)

b. Prolonged Capillary refill time (CRT) - longer than 3 seconds.

c. How to assess CRT - Press gently for 5 seconds and blanch skin over forehead / sternum, release & assess time for refill. (Count 1 in 1000, 2 in 1000, 3 in 1000 in your mind).

d. Persistent tachycardia - HR more than 160 / min. Check HR (resting) for a while on monitor. It is not possible to accurately count manually such high heart rates.

e. Tachypnoea with no retractions, suggestive of metabolic acidosis.

f. Lethargy

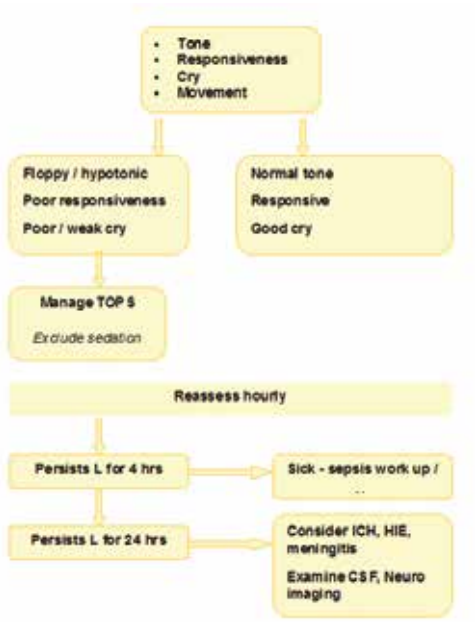
Late signs of poor perfusion (Decompensated shock)

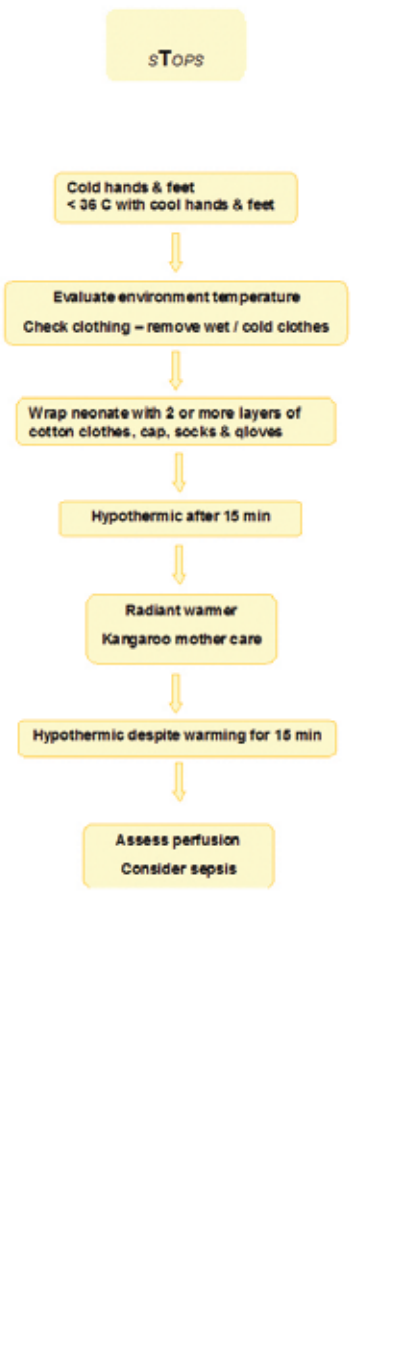
Low BP, poor pulses

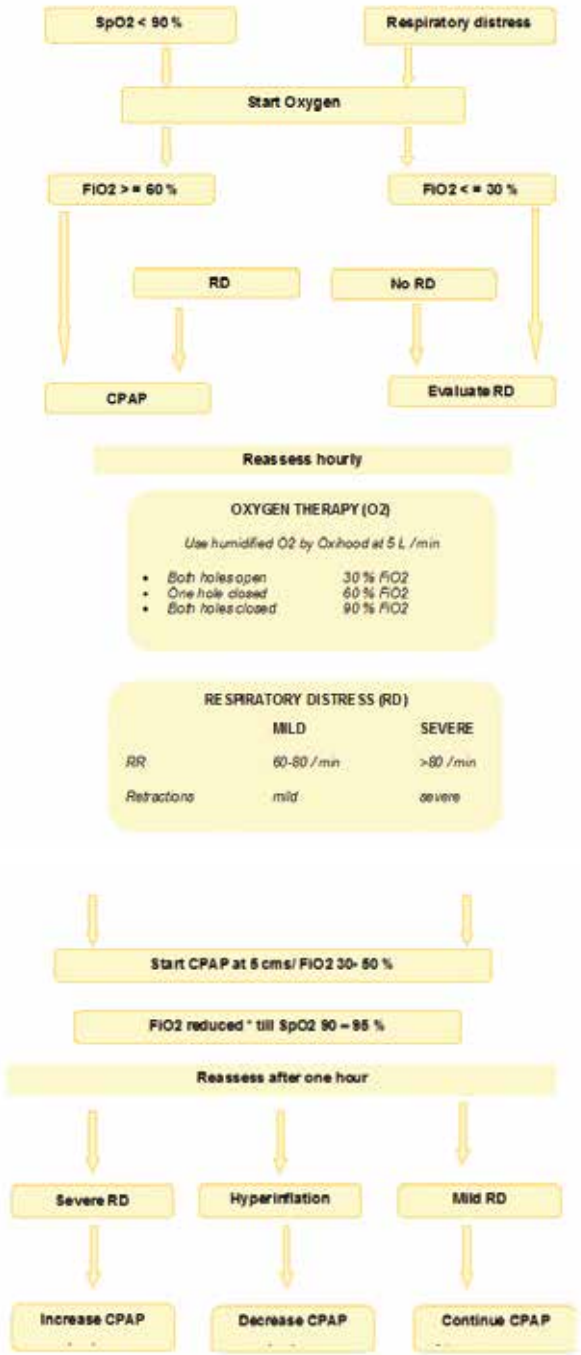
5. SUGARS

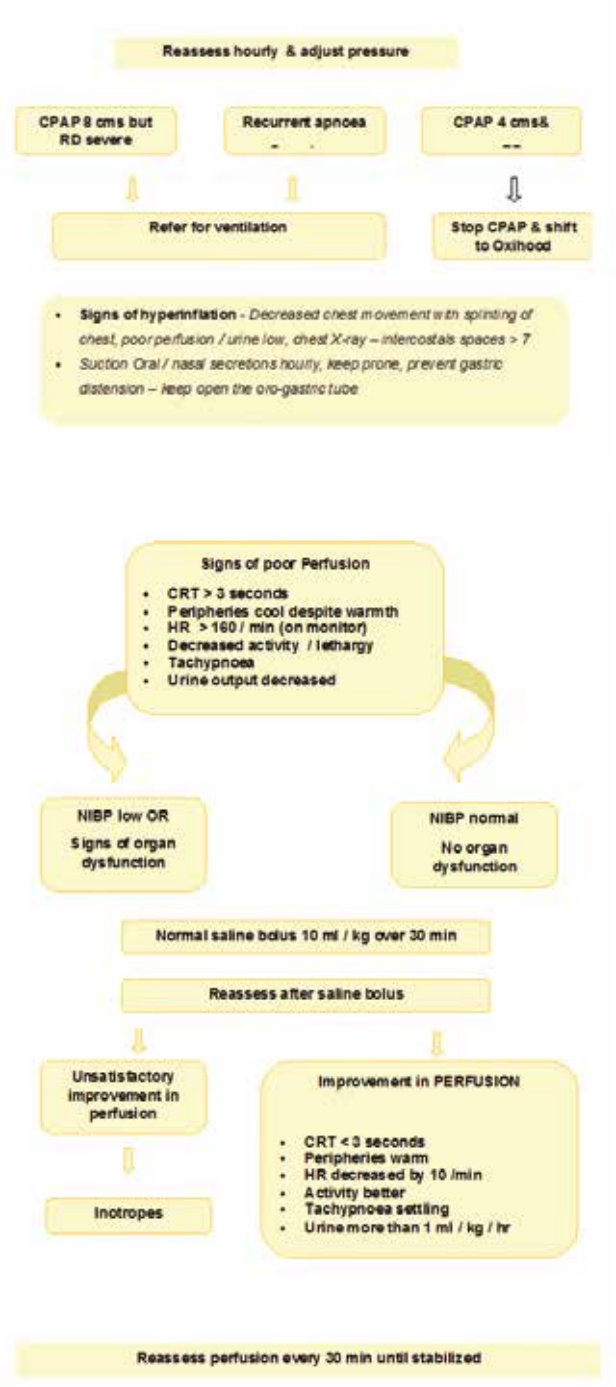
GOOD: Blood BS: 40 - 125 mg / dL

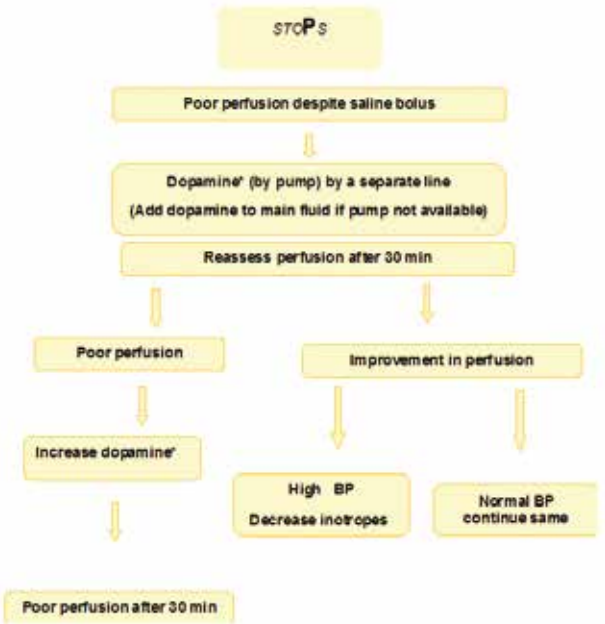
BAD: Low blood sugar (<40) is an emergency, >125 indicates stress

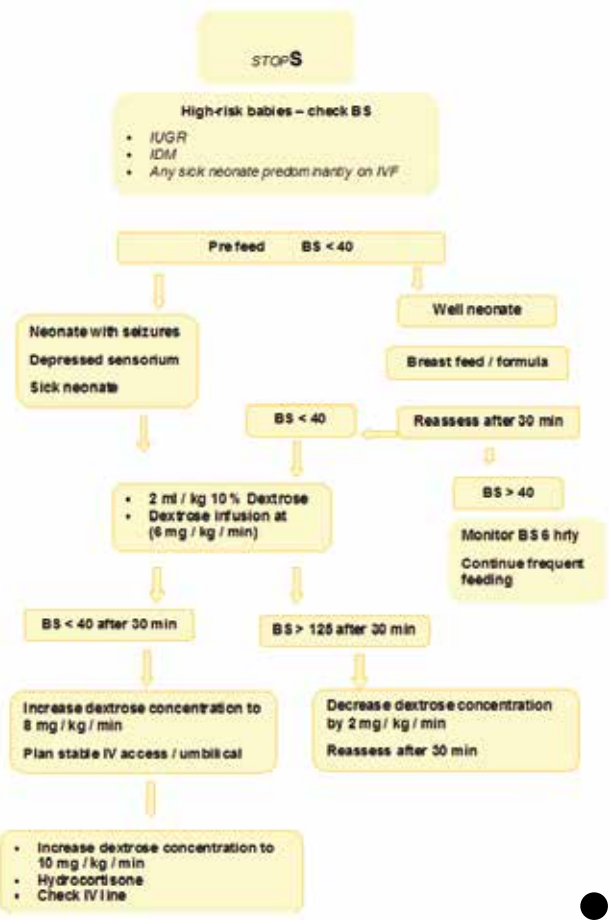



sTOPS
Cold hands & feet
< 36 C with cool hands & feet
Evaluate environment temperature
Check clothing – remove wet / cold clothes
Wrap neonate with 2 or more layers of cotton clothes, cap, socks & gloves
Hypothermic after 15 min
Radiant warmer
Kangaroo mother care
Hypothermic despite warming for 15 min
Assess perfusion
Consider sepsis


SpO2 < 90 %
Respiratory distress
Start Oxygen
FIO2 > = 60 %
FIO2 < = 30 %
RD
No RD
CPAP
Evaluate RD
Reassess hourly
OXYGEN THERAPY (O2)
Use humidified O2 by Oxhood at 5 L /min
Both holes open 30 % FiO2
One hole closed 60 % FiO2
Both holes closed 90 % FiO2
RESPIRATORY DISTRESS (RD)
MILD
SEVERE
RR
60-80 /min
>80 /min
Retractions
mild
severe
Start CPAP at 5 cms/ FIO2 30- 60 %
FiO2 reduced * till SpO2 90 – 95 %
Reassess after one hour
Severe RD
Hyperinflation
Mild RD
Increase CPAP
Decrease CPAP
Continue CPAP


Reassess hourly & adjust pressure
CPAP 8 cms but RD severe
Recurrent apnoea
CPAP 4 cms&
Refer for ventilation
Stop CPAP & shift to Oxihood
Signs of hyperinflation - Decreased chest movement with sprinting of chest, poor perfusion / urine low, chest X-ray – intercostals spaces > 7
Suction Oral / nasal secretions hourly, keep prone, prevent gastric distension – keep open the oro-gastric tube
Signs of poor Perfusion
CRT > 3 seconds
Peripheries cool despite warmth
HR > 160 / min (on monitor)
Decreased activity / lethargy
Tachypnoea
Urine output decreased
NIBP low OR
Signs of organ dysfunction
NIBP normal
No organ dysfunction
Normal saline bolus 10 ml / kg over 30 min
Reassess after saline bolus
Unsatisfactory improvement in perfusion
Inotropes
Improvement in PERFUSION
CRT < 3 seconds
Peripheries warm
HR decreased by 10 /min
Activity better
Tachypnoea settling
Urine more than 1 ml / kg / hr
Reassess perfusion every 30 min until stabilized


STOPS
Poor perfusion despite saline bolus
Dopamine* (by pump) by a separate line
(Add dopamine to main fluid if pump not available)
Reassess perfusion after 30 min
Poor perfusion
Improvement in perfusion
Increase dopamine*
High BP
Decrease inotropes
Normal BP
continue same
Poor perfusion after 30 min


STOPS
High-risk babies – check BS
IUGR
IDM
Any sick neonate predominantly on IVF
Pre feed BS < 40
Neonate with seizures
Depressed sensorium
Sick neonate
Well neonate
Breast feed / formula
BS < 40
Reassess after 30 min
BS > 40
Monitor BS 6 hrly
Continue frequent feeding
2 ml / kg 10 % Dextrose
Dextrose infusion at (6 mg / kg / min)
BS < 40 after 30 min
BS > 125 after 30 min
Increase dextrose concentration to 8 mg / kg / min
Plan stable IV access / umbilical
Decrease dextrose concentration by 2 mg / kg / min
Reassess after 30 min
Increase dextrose concentration to 10 mg / kg / min
Hydrocortisone
Check IV line

Neonatal Cardiac Emergencies

Dr. Remadevi
Consultant Pediatric Cardiologist.
Amrita Institute of Medical Sciences,Kochi

Cardiac emergencies presenting in the neonatal period can be broadly classified in to three based on etiology.

These are,

1. Critical Congenital Heart Disease (CCHD)
2. Primary Myocardial dysfunction (Myocarditis/Cardiomyopathy/Ventricular dysfunction due to other causes like inborn errors of metabolism)
3. Arrhythmias (tachy/bradyarrhythmias)

Critical Congenital Heart Diseases (CCHD)

Definition: CHD requiring surgery or catheter based intervention in the 1st year of life. 25% of CHDs are critical.

CCHDs are the most common cause of a neonatal cardiac emergency.

CCHDs presenting in the neonatal period include belong to two groups

1. Ductus dependent CCHDs

   Duct dependent pulmonary circulation: Pulmonary atresia/ critical pulmonary stenosis with variable intracardiac anatomy.

   Exclusion: Pulmonary atresia with MAPCAs

   Duct dependent systemic circulation: Hypoplastic left heart syndrome, Critical aortic stenosis/atresia, Critical coarctation/Arch interruption with variable intracardiac anatomy.
2. Other CCHDs which are not ductus dependent. Include both cyanotic and acyanotic CCHDs

   Cyanotic: TGA (Open PDA improves saturation in TGA/intact ventricular septum though not ductus dependent), TAPVC, TOF, Truncus arteriosus, Single ventricle

   Acyanotic :Large posttricuspid left to right shunts (Large PDA, Large AP window,Oc-

casionally large VSD).

Timing and symptomatology of presentation depends on two factors.

1. Nature and severity of CHD
2. Changes occurring with transition ie closure of patent ductus arteriosus and fall in pulmonary vascular resistance

Clinical presentation of CCHD

Present in limited number of ways which provide clue to etiology and management

1. Circulatory collapse/shock
2. Cyanosis
3. Cyanosis with Respiratory distress
4. Congestive Heart failure
5. Murmur

Presentation	Underlying CCHD	Clinical findings	Differential diagnosis
Circulatory collapse/shock	Ductus dependent systemic circulation Eg: HLHS, Critical coarctation, Critical aortic stenosis, Aortic arch interruption	Uneventful early postnatal period. Presents in shock when PDA constricts. Other features of heart failure like gallop rhythm and hepatomegaly. Absent/reduced pulses in all extremities/lower limbs depending on etiology, Minimal clinical cyanosis(differential cyanosis with UL-LL difference >3% may be present in coarctation/interruption) CXR: Cardiomegaly with pulmonary congestion	Myocardial dysfunction due to varying causes(Myocarditis, Inborn errors of metabolism), Septic shock
Cyanosis	Ductus dependent pulmonary circulation-DDPC(Eg: Pulmonary atresia irrespective of the intracardiac anatomy, Critical pulmonary stenosis, Neonatal Ebsteins anomaly) and TGA with intact ventricular septum	Presents with worsening cyanosis and metabolic acidosis when PDA constricts. Normal early postnatal period(unless pulse oxymetry picked up subclinical cyanosis). Tachypnoea occurs to compensate for metabolic acidosis and hence manifests late. Systemic perfusion and BP are maintained till the very end. CXR: No significant Cardiomegaly(exception: Neonatal Ebsteins anomaly which presents with massive Cardiomegaly) Pulmonary blood flow: Normal(in TGA) /reduced(relatively black lungs in DDPC)	Primary lung disease CNS abnormailities Methhemoglobinemia(Normal PO2 in ABG)
Cyanosis with respiratory distress	Eg: Obstructed TAPVC (Isolated or associated with complex CHD like heterotaxy syndrome). Rarely HLHS with restrictive interatrial septal opening.	Worsening respiratory distress with variable cyanosis within an hour to days after birth. May present immediately after birth. Often requires ventilation and even HFOV. Perfusion maintained in the initial stages. CXR: Hazy lung fields due to pulmonary oedema caused by pulmonary venous obstruction (Variable degree of white out). No significant Cardiomegaly.	Other causes of Neonatal RDS and PPHN like MAS, HMD, and Malignant TTNB *(All newborns with diagnosis of PPHN, lower PO2 with high ventilator requirements should have a suspicion of obstructed TAPVC, even if one hasty echo diagnoses only PPHN)*
Congestive heart failure	1. Admixture lesions (Complete mixing of oxygenated & deoxygenated blood). Eg: Unobstructed TAPVC, Single ventricle,Truncus arteriosus, 2.Large left to right shunts: Large VSD(less likely), PDA, AP window, Complete AV canal defect	Insidious CHF within days to weeks after birth. Cyanosis present in admixture lesions, but minimal (Often detected by saturation monitoring). Perfusion and pulses usually preserved till late(High volume pulses in PDA, Truncus and AP window). CXR: Shows Cardiomegaly and pulmonary plethora.	Heart failure due to myocardial dysfunction(Myocarditis, Inborn errors of metabolism), Large Arteriovenous malformations, Severe anemia.
Murmur	At birth: Tetralogy of Fallot and other CHDs having tetralogy physiology, Isolated outflow tract stenosis(AS/PS), Within days to weeks: Posttricuspid left to right shunts like VSD/PDA	Depends on the CHD. Many critical CHDs present with no murmur. Hence murmur is not a dependable finding. When present, needs evaluation for other features of CCHD and echocardiogram should follow.	

Diagnosis and management

Initial stabilization based the clinical features precedes anatomical diagnosis.

Hyperoxia test

Should ideally be performed in all neonates with suspected critical CHD (Not only in those who are cyanotic),unless there is immediate access to echocardiography performed by an experienced person. It is a sensitive and specific tool to differentiate CCHD from pulmonary disease.

Arterial oxygen tension is measured in the right radial artery (preductal) in room air and while the patient breathes 100 percent oxygen(via a hood or endotracheal tube if baby is ventilated) for 10 minutes.. A significant increase in systemic arterial oxygen saturation and partial pressure of arterial oxygen (PaO2) above 150 mmHg during the hyperoxia test makes it more likely that the patient has lung disease and not CCHD.

However, exceptions are there. These are

1. Failure to increase PaO2 >150mm Hg may occur occasionally with severe lung disease with PPHN too.
2. In admixture lesions with increased pulmonary blood flow(eg: Truncus arteriosus, HLHS), rarely PaO2 >150mm Hg may be obtained with hyperoxic challenge(because of significantly increased pulmonary blood flow as pulmonary vascular resistance falls with O2)

*Pulse oxymeter should not be used for hyperoxia testing. Because, normal haemoglobin is fully saturated with oxygen when the arterial PO2 exceeds 70 mmHg. Hence it will not detect inadequate increase in arterial PO2.

General supportive measures:

Based on principles of neonatal advanced life support.

Airway and ventilation to be ensured. Reliable vascular access to be obtained with stress on volume resuscitation, inotropic supports and correction of metabolic acidosis, as the condition demands.

Use of supplemental oxygen:

Patients with hypoxia due primary lung pathology or cardiogenic pulmonary oedema (obstructed TAPVC, HLHS with restrictive ASD) are likely to benefit from use of supplemental oxygen or higher FiO2.

Higher inspired oxygen concentration is harmful in neonates with ductus dependent systemic circulation like HLHS and hence should be avoided.

Hypercarbia and hypocarbia:

Some amount of hypercarbia with resultant mild respiratory acidosis (pH: 7.35) is beneficial in HLHS, increasing pulmonary vascular resistance and improving the systemic oxygen delivery.

Hypercarbia may precipitate a pulmonary hypertensive crisis with acute worsening of cardiovascular status in case of obstructed TAPVC.

Hence , it is important to maintain normal PCO2 levels when dealing with an undiagnosed CCHD.

Specific measures

1. Prostaglandin E1 (PGE1, Alprostadil)

Indication:

1. Neonates presenting in a critical ill condition within the first 2 to 3 weeks of life with cyanosis or shock where duct dependent CHD or TGA is strongly suspected. Echo confirmation is not mandatory.

   It should be continued till duct dependent CHD is ruled out after echocardiogram or definitive treatment is done in case of a duct dependent CHD.

2. Antenatally diagnosed newborns with ductus dependent CHD should be started on PG before they become critically ill.

   Ductus dependent pulmonary circulation: PG may be started when saturation falls below 75 to 80%

   Ductus dependent systemic circulation: It is better to start PG in low dose soon after birth as early signs of constriction of PDA like pulse discrepancy in coarctation, are more subjective.

Dose and administration

Given as continuous intravenous infusion. Peripheral venous access is adequate. Additional venous access is advised for other medications and fluids.

Dose: Depends upon the clinical scenario. Aim is ductal patency at lowest effective dose.

- When we start PG in a stable newborn with previously diagnosed ductus dependent circulation, starting dose of 0.01mcg/kg/min is enough.
- In criticall ill Newborn with constricted PDA, higher dose of 0.05mcg/kg/min is required to open up the PDA. This is the standard starting dose.
- Dose may increased up to 0.1mcg/kg/min if needed
- Store between 2 to 8 degree C.
- Use freshly prepared infusion solutions. Discard any unused solution kept for more than 24hours.
- Method of administration: Usually mixed in 5% dextrose. Each vial contains Alprostadil 500mcg/1ml. 0.5ml (250mcg) of PGE1 is diluted with 50ml of 5% dextrose.

  Infusion rate in ml/hr= (dose of PGE1 in mcg/kg/min x wt in kg x 50 x60) ÷ 250

  Reassess 15 to 30 min after starting PGE1. Once necessary effect (Saturation > 80% in ductus dependent pulmonary circulation and good lower limb pulses and BP with correction of acidosis in ductus dependent systemic circulation) is achieved, dose may be tapered up to 0.01mcg/kg/min.
- Adverse effects.

  Two major dose dependent side effects are respiratory depression and hypotension. Using the lowest effective dose minimizes both. Apnea occurs in 10 to 12% usually within the 1st 6hrs of starting the infusion. Hypotension with tachycardia occurs due to vasodilatation and responds to volume resuscitation.

Conditions worsened by PGE1

- Obstructed TAPVC and Other conditions with pulmonary venous obstruction like HLHS/mitral atresia with restrictive ASD. It is said that PDA may increase pulmonary oedema. However, this fear often theoretical, because PDA shunts right to left in such conditions and hence unlikely to worsen the situation.
- TGA with very restrictive ASD

  These conditions can be made out by CXR ie. Hazy lungs(indicating pulmonary oedema) with disproportionate respiratory distress usually indicates pulmonary venous congestion.

2. Echocardiogram

Primary diagnostic tool based on which management plan is to be made. Hence arrangements should be made during initial stabilization. Once echocardiographic confirmation is obtained, treatment and stabilization may be modified accordingly.

3. Transport

Once stabilized and basic anatomic diagnosis is made by echocardiogram, neonate needs to undergo definitive lesion specific treatment. This may require transfer to an institution with pediatric cardiac programme.

Care during transport

- Neonates receiving high dose PGE1 should be intubated and transported (for fear of apnea). However, if the baby is on low dose PGE1 and observed for 4 to 6 hrs before transport, ventilation may be avoided.
- Reliable vascular access should be secured. Umbilical lines if secured should be left in place.
- Arterial blood gas to be obtained and acid base status and oxygen delivery optimized especially in ventilated newborns.
- Supplementary oxygen and FIO2 should be adjusted based the specific diagnosis (as discussed in lesion specific care)
- Consultation with pediatric cardiologist at the receiving centre helps in optimising transport status.

- Coordination between referring team, transport team and receiving team on all aspects of patient care is important.

3. Lesion Specific Care / Definitive treatment for neonates with symptomatic CCHD

Majority of critical CHDs can treated by curative or palliative interventions with good outcome.

Treatment can either be percutaneous transcatheter intervention or surgery depending on the lesion.

Ductus dependent pulmonary circulation	
Critical Valvar Aortic stenosis	Percutaneous Transcatheter Balloon Aortic valvotomy
Critical coarctation of aorta/ Interruption of aortic arch:	Surgical repair
Hypoplastic left heart syndrome	Excessive supplemental oxygen results in pulmonary vasodilation with excessive pulmonary blood flow with resultant systemic underperfusion and metabolic acidosis. Hence, ventilating in room air will be optimal, provided there is no additional lung issue. Saturation of 80% with normal
	pH indicates acceptable hemodynamic status. Definitive treatment is staged surgical palliation including a high risk, neonatal Norwood procedure or neonatal cardiac transplantation.
Duct dependent pulmonary circulation:	
Critical valvar pulmonary stenosis	Percutaneous transcatheter balloon pulmonary valvotomy.
Valvar Pulmonary atresia with intact ventricular septum:	Needs transcatheter(preferred) or surgical relief of RV outflow obstruction with or without an aorto pulmonary shunt or PDA stenting. In case of very hypoplastic RV, only palliation(BT shunt/PDA stenting is done).
VSD or single ventricle with pulmonary atresia/critical pulmonary stenosis	Needs staged intervention. 1st step in neonatal period is a palliative procedure to ensure a reliable source of pulmonary blood flow (Palliative BT shunt or PDA stenting). Further surgical interventions needs to be done later, depending on anatomy.
Transposition of great arteries (Parallel circulation)	Saturation depends on mixing between systemic and pulmonary circulations (via ASD, PDA and, VSD if present). TGA with intact ventricular septum presents with hypoxemia within hours after birth, when PDA constricts. As most newborns with have a PFO, PG E1 is often helpful to maintain saturation above 70% till intervention is done. If PFO is very small and immediate arterial switch operation is not possible, balloon atrial septostomy(BAS) may be done to improve mixing. PG E1 may be started, if BAS alone fails to improve saturation adequately. Corrective surgery (arterial switch operation) should be done within days to weeks of life.
Lesions with complete intracardiac mixing	
Total anomalous pulmonary venous connection	Obstructed TAPVC is a surgical emergency where PG E1 do not have any role and medical stabilization only has limited role. However, optimal ventilation(even requiring HFOV) , inotropes and sedation will help to tide over the crisis till urgent surgery is made available. Unobstructed TAPVC presents with less severe symptoms and requires early elective surgical correction.
Truncus arteriosus	Early surgical correction. Medical management of heart failure(diuretics, digoxin) till surgery is done
Left to right shunts(Large PDA, AP window, VSD, Complete AV canal defect)	Medical stablilization with diuretics and digoxin. Surgical correction at 2 to 4months of age or earlier if symptomatic.

Primary myocardial dysfunction

Viral myocarditis is the commonest etiology (Common viruses: Enteroviruses particularly Cocksackie virus, Adenovirus, parvovirus B 19 etc). Diagnosis is mostly presumptive after excluding other causes like inborn errors of metabolism.Presents with congestive heart failure/cardiogenic shock or arrhythmia. Can be fatal. Partial or complete recovery occurs in those survive the acute stage.

Management

Supportive care is the main stay. This includes inotropic support,diuretics and mechanical ventilation as required.

Diuretics: Provide earliest symptomatic relief provided perfusion is adequate

Inotropic support: Drugs like dobutamine and milrinone which donot increase systemic vascular resistance are preferred.

Mechanical ventilation: Takes away the work of breathing and treats pulmonary oedema

IVIG: Not supported by evidence and hence not advised.

Temporary mechanical circulatory support (ECMO) may be required in fulminant cases till myocardium recovers.

Arrhythmias

One of the common cardiac emergencies in newborns. ECG is essential for accurate electrophysiological diagnosis and management. ECG to be analysed for rate, rhythm and QRS morphology.

Broadly categorized in to tachyarrhythmia and bradyarrhythmia. Tachyarrhythmia: Can be narrow QRS or wide QRS tachycardia. Wide QRS tachycardia is uncommon in neonates.

Narrow QRS tachycardia

- Usually of supraventricular origin.
- Most common symptomatic arrhythmia in children including neonates.
- Accessory pathway mediated re-entry tachycardia is the commonest mechanism (Atrioventricular reciprocating tachycardia-AVRT) followed by atrial flutter.
- ECG features of AVRT: Rate >200/min(often as high as 240 to 300/min in newborns and young infants), regular with inverted P seen on ST segment. Causes heart failure when sustained for many hours. Usually occurs in structurally normal hearts.

Treatment

If in circulatory collapse- Administer synchronized DC cardioversion at 0.5-1.0 J/kg - if not effective increase to 2 joules/kg .

If relatively stable: Adenosine is the drug of choice. Get an ECG and give adenosine 100mcg/kg to 200mcg/kg rapid IV push in to a large vein (antecubital) followed saline bolus push to drive it in to heart.

Adenosine has short half-life of <10 seconds. Maximum dose is 12mg or 400mcg/kg/dose.

Diagnostic use of adenosine

In case of atrial flutter, 2nd most common type of SVT, there is acute AV block with unmasking of sawtoothed flutter waves. Synchronized DC cardioversion is the effective treatment for atrial flutter.

ECG is a must during adenosine administration to interpret response or nonresponse.

Facility for DC cardioversion should be ready while administering

adenosine, as it may precipitate atrial fibrillation.

Reason for 'why adenosine didn't work'

1. Non AV node dependent arrhythmia like atrial flutter.
2. Slow administration /inadequate dose so that drug could not act: Rpt dose at 200mcg/kg with proper precautions to be given.
3. Transient termination with a few sinus beats followed by recurrence of arrhythmia.
4. Problems with potency of drug. A different batch may be tried.

If the drug has not produced transient AV block, that means drug has not acted.

After restoring sinus rhythm, maintenance therapy should be started to prevent recurrence. Betablockers like propanolol are the mainstay.

Other medications

Amiodarone:

Loading with amiodarone is helpful in refractory SVTs. Dose 5 mg/kg as intravenous infusion over 20 to 60 minutes. Can be repeated up to a total loading dose of 15mg/kg/min. Beware of acute cardiovascular collapse due to hypotension caused by its acute alpha receptor blocking effects particularly in unstable patients and neonates.

Indication in the treatment of SVT include

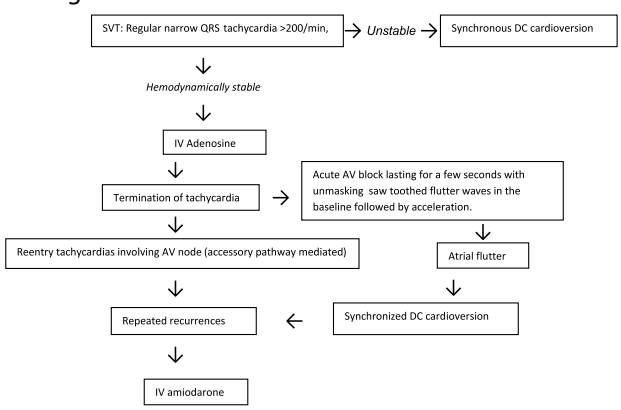
1. Repeated recurrences despite betablockers to prevent recurrence, in case of AVRT and atrial flutter
2. Other uncommon tachycardias not responding to adenosine and which can cause tachycardiomyopathy.eg: Atrial tachycardia, PJRT.

Multiple adverse effects limits its use. Hence, it should be used only when indicated and for the shortest duration.

Flecainide:

It is used to control supraventricular tachycardias in structurally normal hearts when recurrences of arrhythmia is not controlled by betablockers alone.

Management of SVT



Bradyarrhythmias

Commonest etiology: Congenital Complete Atrioventricular block(CCHB). May occur associated with structural heart disease or maternal autoantibodies which are transplacentally transmitted(anti SS-A/Ro and/ or anti SS-B/La). ECG is diagnostic. Presence or absence of symptoms depend on patient's ventricular escape rate and rhythm.

Treatment

Definitive treatment is permanent pacemaker implantation(PPI). Temporary Pacing via transvenous route can be achieved till PPI is done.

Indications PPI in infancy include

1. Symptoms, ventricular dysfunction or low cardiac output
2. Wide QRS escape rhythm and complex ventricular ectopy
3. Ventricular rate (VR) <55/min in those with structurally normal heart and VR <70/min in those with CHD, Pauses > 2 to 3 times the basic cycle length in holter monitoring.

Asymptomatic newborns and infants not having these indications are kept on follow up.

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Approach to HYPOTENSION in Neonates

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In newborns, blood pressure (BP) varies with gestational age, postmenstrual age, and birthweight. BP increases after birth, with greater rates of increase seen in preterm infants than in term infants. Recent studies have shown that there is significant variability within the extremely low-birthweight (ELBW) population both in measurements and responses to therapies for hypotension.

DEFINITION OF HYPOTENSION

There is no standard definition of hypotension in neonates. In clinical trials and in practice, hypotension is defined as any value that falls below the 5^{th} or 10^{th} percentile for gestational and postnatal age. The most accepted definition of physiologic hypotension is the point at which cerebrovascular autoregulation is lost, leading to cerebral function compromise and tissue ischemia.

MEASUREMENT OF BLOOD PRESSURE IN NEONATES

The "gold standard" for determining BP in the critically ill neonate is a direct reading from an indwelling arterial line, and should be used whenever arterial access is available.

MBP is considered most reflective of the systemic perfusionpressure because the systolic and diastolic values arethought to be affected by the small bubbles that may getintroduced in the system. Other non-invasive methods ofmonitoring include the use of Doppler or oscillometrictechniques, but their inability to provide continuous monitoring is a major drawback.

Mean Blood Pressure (mm Hg) in Neonates With Gestational Ages 23 Weeks To Term*

	Postnatal age (hours)						
Gestational age (weeks)	0	12	24	36	48	60	72
23–26	24	25	26	27	28	29	30
27–32	30	31	32	33	34	35	36
33–36	36	37	38	39	40	41	42
≥37	43	44	45	46	47	48	49

*[illegible]

Physiology:

Principal factors governing circulatory function

1. Preload
2. Inotropy
3. Afterload

Etiopathology:

In term babies:-

- Hypovolemia:-
  a. Hemorrhagic:- Antepartum /post partum losses
  b. Non hemorrhagic:- fluid and electrolyte losses
- Cardiogenic:-
  a. Cardiac:- Congenital heart disease, Cardiomyopathies, Arrhythmias, PDA in preterm
  b. Secondary cardiac:- Birth asphyxia, Sepsis,PAH, IEM
- Distributive:- Adrenal insufficiency (Congenital adrenal hyperplasia, adrenal hemorrhage), septic, neurogenic.
- Obstructive:- Tension pneumothorax, pericardial tamponade

Factors Contributing To Hypotension In Preterm Neonates

1	Immature myocardium → decreased contractility
2	Transition from fetal to perinatal circulation → increased SVR
3	PDA → left-to-right shunt → steal syndrome
4	Perinatal hypoxia/asphyxia → neuroendocrine changes causing increased SVR
5	PPV → decreased venous return
6	Sepsis and inflammation → inflammatory mediators causing peripheral vasodilation and increased vascular permeability
7	Relative adrenal insufficiency → insufficient cortisol during stress/illness

PDA=patent ductus arteriosus; PPV=positive pressure ventilation; SVR=systemic vascular resistance.

ADVERSE EFFECTS OF HYPOTENSION IN NEONATES

The ultimate aim of maintaining adequate blood-pressure is to ensure satisfactory tissue perfusion. This has proved difficult to measure. Some small studies have suggested white matter damage and poor neuro-development outcome associated with recorded hypotension (variably defined), however, no such correlation was observed in a large cohort of VLBW infants.

Assessment and Management

A careful clinical and biochemical assessment of a potentially hypotensive infant is an essential first step towards management. This should include: heart rate, capillary refill time, urine output, serum lactate concentration, pH, base excess and haemoglobin. A conservative approach (permissive hypotension) is acceptable if the clinical examination is satisfactory in the face of apparent hypotension.

20mls/kg fluid bolus (0.9% saline over 30-60min). Consideration should be given for the need for blood products (including FFP if clotting deranged). Assess need for second bolus if remains clinically hypovolaemic. This may be justified as infants rarely reach the peak of the Frank-Starling curve.

⇩

Start dopamine at 6-10 mcg/kg/min (dose range 2-20 mcg/kg/min). If no response, consider asking for cardiology review and Echo, to help asses filling and ventricular function.

⇩

Consider Dobutamine. Start at 6-10mcg/kg/min (dose range 2-20 mcg/kg/min). Dobutamine may be inappropriate in profound vasodilatation.

⇩

Consider starting Hydrocortisone (2.5mg/kg/dose 6hrly). Recent evidence shows that a smaller dose of 2mg/kg/day may also be equally effective

⇩

Consider Adrenaline 0.05- 0.3 mcg/kg/min

(Choice of treatment of low SVR with or without impaired contractility (septic shock)

In all other situations, when unresponsive to high dose dopamine)

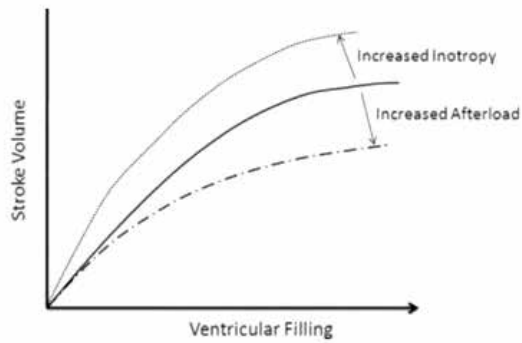
Vasopressin:

- In catecholamine resistant hypotension in vasodilatory shock.
- No large RCTs yet

Milrinone:

- Near term and term neonates with PAH
- Increasingly used to treat low cardiac output after corrective cardiac surgeries

Frank-starling curve showing effect of increased inotropy versus increased afterload on stroke volume



Cochrane reviews

- Dopamine was more successful than albumin at correcting low BP in hypotensive preterm infants (Osborn and Evans 2001).
- Dopamine is more effective than dobutamine in the short term treatment of systemic hypotension in preterm infants (Subhedar and Shaw 2003).
- There are insufficient data on the use of adrenaline infusions in preterm infants with cardiovascular compromise (Paradisis and Osborn 2004).
- Hydrocortisone may be as effective as dopamine when used as a primary treatment for hypotension (Ibrahim, Sinha, and Subhedar 2011)

Adrenergic and dopaminergic receptor-dependent cardiovascular actions of the most frequently used sympathomimetic agents

Agent	Cardiovascular adrenergic and dopaminergic receptors*					
	Cardiac receptors			Peripheral vascular receptors		
	α_1↑	β_1 (β_2)↑	Dopamine	α_1/α_2	β_2	Dopamine
	↑contractility	↑rate ↑conduction ↑contractility	↑contractility	Peripheral vaso-constriction	Peripheral vasodilation	Vasodilation in renal, mesenteric, and coronary circulation
Dopamine[#]	++[#]	++++[#]	+	++++	+	++++
Epinephrine	++	++++	Ø	++++	+++	Ø
Norepinephrine	++	++++	Ø	++++	Ø/+	Ø
Dobutamine§	++	++++	Ø	+	++	Ø

Estimated relative contribution of adrenergic and dopaminergic receptor stimulation to the cardiovascular actions of sympathomimetic amines.
Ø no effect; + minimum effect; ++++ maximum effect.*, receptors other than the adrenergic and dopaminergic receptors also mediate some of the cardiovascular actions of the sympathomimetic agents (for example, the dopamine-induced stimulation of the serotonin receptors contributes to the peripheral vasoconstriction caused by higher doses of the drugs).#, dopamine stimulates the α_1- and β-adrenergic and dopaminergic receptors in a dose-dependent manner. §, only approximately 10% of the positive inotropic effect of dopamine result from the direct stimulation of the myocardial adrenergic receptors [9]. ¶, the relative contribution of the α_1- and β_1-adrenergic receptors and the myocardial dopamine receptors to the increase in myocardial contractility in the neonate is unknown. ¥, 3-O-methyldobutamine, the major dobutamine metabolite, is a relatively potent and highly selective α_1-adrenoceptor antagonist [10]. The α_1-adrenoceptor inhibitory effects of this metabolite may contribute to the tendency of dobutamine to cause peripheral vasodilation. See text for details.

Neurological Emrgencies in Neonate

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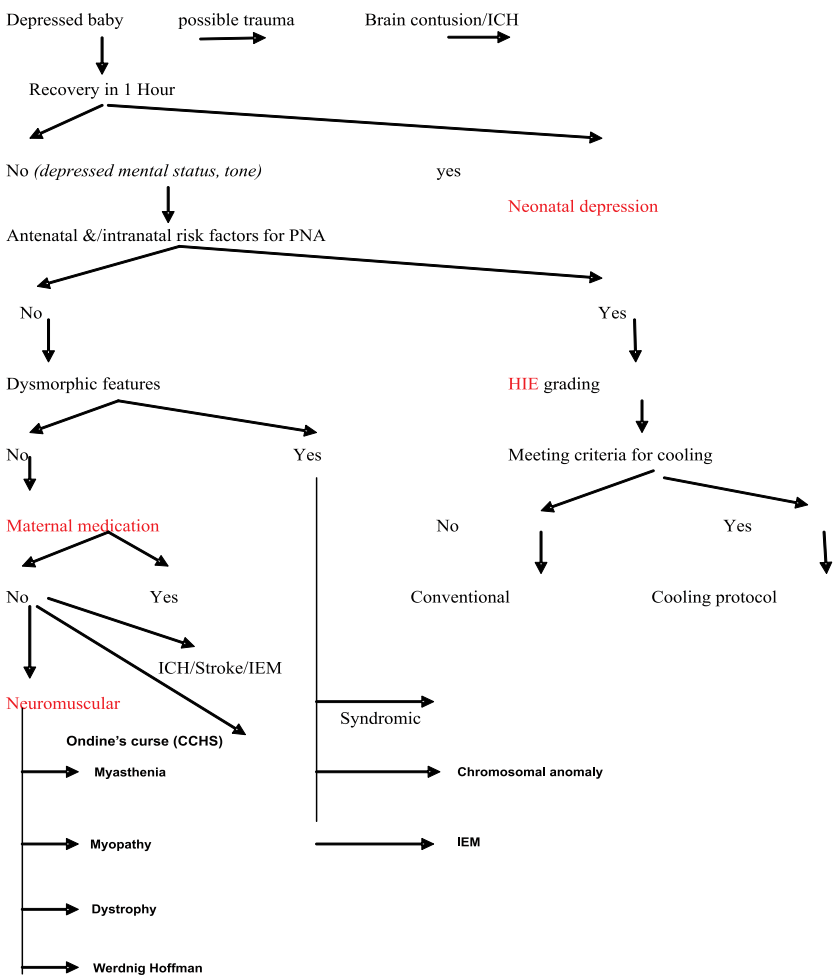

neonatal depression:a descriptive term of the condition of the newly born infant in the first hour of birth, includes depressed mental status, muscle hypotonia, and/or disturbances in respiration and circulation.

Assessment of neonatal encephalopathy (NE) by National Institute of Child Health and Human Development (NICHD) score

	NORMAL	MILD NE	MODERATE NE	SEVERE NE
1. Level of consciousness	1. Level of consciousness Alert, Responsive to external stimuli (state dependent, eg. post feeds)	Hyper-alert, has a stare, jitteriness, high-pitched cry, exaggerated responds to minimal stimuli, inconsolable	Lethargic	Stupor/coma
2. Spontaneous activity	Changes position when awake	Normal or Decreased	Decreased activity	No activity
3. Posture	Predominantly flexed when quiet	Mild flexion of distal joints (fingers, wrist usually)	Moderate flexion of distal joint, Complete extension	Decerebrate
4. Tone	Strong flexor tone in all extremities + strong flexor hip tone	Normal or Slightly increased peripheral tone	Hypotonia (focal or general) or Hypertonia	Flaccid Rigid
5. Primitive reflexes (Circle only the highest level in each sign; The maximum score is only one in any one category)	Suck Strong,	Weak, poor	Weak but has a bite	Absent
Moro	Complete	Partial response	Low threshold to elicit	Incomplete Absent

TOTAL SCORE

* Seizure None None Yes / No Yes / No

Infant who has seizure will be Moderate or Severe NE depending on the neurologic exam. Seizure with normal or mild NE or moderate NE on neurologic exam will be “Moderate NE”. Seizure with severe NE will be “Severe NE”. The level of encephalopathy will be assigned based on which level of signs (moderate or severe) predominates among the 6 categories. If moderate and severe signs are equally distributed, the designation is then based on the highest level in Category #1: The level of consciousness. If the level of consciousness is equal, then allocate the NE stage based on the tone.

Levene’s encephalopathy grading-condone seizure(con…tone….sei…..su….re)

Feature	Mild	Moderate	Severe
Conscious	Irritable	Lethargy	Comatose
Tone	Hypotonia	Marked	Severe
Seizures	No	Yes	Prolonged
Sucking / respiration	Poor suck	Unable to suck	Unable to sustain spontaneous respiration

Whole body cooling

Inclusion criteria.

a. Postmenstrual age (PMA) ?36 weeks, BW ?2,000 g
b. Evidence of fetal distress or neonatal distress as evidenced by one of the following:
    i. History of acute perinatal event (e.g., placental abruption, cord prolapse, severe FHR abnormality)
    ii. pH ?7.0 or base deficit ?16 mmol/L in cord gas or postnatal blood gas obtained within first hour of life
    iii. 10-minute Apgar score of ?5
    iv. Assisted ventilation initiated at birth and continued for at least 10 minutes
c. Evidence of moderate to severe neonatal encephalopathy by exam and/or aEEG as follows:
    i. Primary method for determining neonatal encephalopathy is physical exam.
    ii. If exam shows moderate or severe encephalopathy, aEEG should be performed to provide further assessment and monitoring.
    iii. In circumstances in which physical exam is unreliable (e.g., muscle relaxants), an aEEG should be performed to determine if there is encephalopathy.
    iv. Patterns on aEEG that indicate moderate or severe encephalopathy include the following,
        a. Severely abnormal: upper margin <10 ?V
        b. Moderately abnormal: upper margin >10 ?V and lower margin <5 ?V
        c. Seizures identified by aEEG

Note: A normal neurologic exam does not require confirmation by aEEG.

2. Exclusion criteria.
    a. Presence of lethal chromosomal abnormality (e.g., trisomy 13 or 18)
    b. Presence of severe congenital anomalies (e.g., complex cyanotic congenital heart disease, major CNSanomaly)
    c. Symptomatic systemic congenital viral infection (e.g., hepatosplenomegaly, microcephaly)
    d. Symptomatic systemic congenital bacterial infection (e.g., meningitis, DIC)
    e. Significant bleeding diathesis
    f. Major intracranial hemorrhage

Approach to neonatal seizure

First 24 Hrs

Soon after birth

HIE

Cerebral contusion

Accidental injection of lignocaine

Later

HIE

Intracranial haemorrhage

Early hypoglycaemia

Early hypocalcemia

Non ketotic hyperglycinemia

Hyperammonemia conditions

Pyridoxine dependency

1 to 3days

Intracranial haemorrhage

Hypoglycaemia, Early hypocalcemia

IEM(Organic acidemias, MSUD, Urea cycle disorders etc)

Familial benign neonatal seizure

Narcotic withdrawal

4 to 7 days

Meningitis

"TORCH"

Brain malformations

Bilirubin encephalopathy

Indigenous "medicines"

5th day seizure

Beyond 1 week

Meningitis

IEM

Intracranial haemorrhage

Cerebral dysgenesis

Late hypocalcemia

Epileptic syndromes

History

Pregnancy/labour history:

- history suggesting possible perinatal hypoxic insult
- Fetal brain abnormalities on antenatal imaging
- HELLP syndrome, particularly if associated with acute fatty liver infiltration, may indicate long chain 3 hydroxyacyl-coenzyme A dehydrogenase (LCHAD) deficiency2
- Maternal medication esp. anticonvulscants/illicit drug use
- history suggesting possible "TORCH"infection
- GDM
- History suggesting trauma, accidental/ non accidental falls or road traffic accident, and inflicted (assault)

Maternal past obstetric history:

- Abortions, stillbirths or neonatal deaths(genetic, thrombophilia and metabolic causes)

Maternal past medical history:

Diabetes

- history suggesting possible of thrombophilia or clotting disorder(DVT etc)
- history suggesting myasthenia, myotonic dystrophy which may lead to secondary HIE
- Cataracts: may indicate inborn error of metabolism, myotonic dystrophy, COL4A1 mutations
- Stiffness or startling: consider myotonic disorders or hyperekplexia
- If muscle aches, pains and tetany exist, consider maternal hyperparathyroidism
- Features of autoimmune disorder

Family history

Family history of neonatal seizures/mental retardation/seizure disorder eg.TS, benign familial neonatal seizures

Consanguinity (IEM)

Family history of neuromuscular disorder eg. Myotonic dystrophy

Siblings with "cerebral palsy": suggestive of vascular abnormalities, such as COL4A1 gene mutations, or thrombophilia

Examination of the parents is important where a neuromuscular disorder is suspected.

Examination of the neonate suggesting possible metabolic cause:

- Abnormal odour of urine
- Skin rash
- Dysmorphism including

Genital abnormalities

Abnormal, inverted nipples

Abnormal fat pads

Abnormal head size/AF

- Liver involvement

Hepatomegaly/ Jaundice

- cardiomyopathy
- Eye abnormalities

Cataracts,Retinitis pigmentosa, Cherry red spots, Optic atrophy, Lens dislocation

- Abnormal odour of urine

First line investigations:

Full blood count: infection, haemorrhage, thrombocytopenia.

PT, APTT: coagulation disorders and intracranial haemorrhage.

Direct Coombs test CSF study including Glycine, L/P ratio if clinically indicated

Liver function test : bilirubin encephalopathy, metabolic conditions, infections

Blood glucose, Urea and electrolytes :

ABG with Blood lactate: A persistently high lactate should trigger further investigations.

aEEG using a cerebral function monitor for identifying and monitoring seizures .

Second line investigations(available quickly)if suspecting non HIE cause for seizure/encephalopathy)

Urinary ketones :presence suggests a metabolic disorder.

Ammonia. Very high levels (>200 ?mol/L) suggests a metabolic cause(urea cycle defect,organic academia)

TMS Urine GCMS

MRI studies

Clinical clues to etiological diagnosis of seizure

Clonic seizures on 2/3rd day with alert inter ictal period in a term baby suggests sub arachnoid haemorrhage.

Hyperactive startle,generalised myoclonus,hypertonia with positive family history-Hyperekpiexia

Purely clonic seizures onset on day 4 to 7 with out any family history in a term baby-5th day seizure

Tonic clonic seizures with family history suggesting dominant inheritance onset on day 2 to 4 Benign familial neonatal seizure

myoclonic seizures and hiccups -NKH

Opisthotonic posturing alternating with flaccidity-MSUD.

Approach to intra cranial bleed in newborn

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When to suspect?

- Predisposing factors- prematurity, trauma, HIE
- Sudden unexplained pallor or jaundice
- Bulging AF
- Refractory Seizures
- Gross Hypotonia/encephalopathy

5 Common major type of bleeds

- GMH/ Intraventricular- common, preterm, serious
- Primary Subarachnoid- common ,both in preterm and term, benign
- Subdural - Uncommon, term infants, serious
- Cerebellar- uncommon, more in preterm, serious
- Intracerebral- uncommon, term, variable severity

Clues from history and examination

- Preterm- IVH, cerebellar
- Large baby, traumatic delivery, breech- subdural
- Well baby with seizures on 2nd or 3rd day- primary subarachnoid
- Perinatal asphyxia- any type of bleed , subdural rare
- Bloody CSF- IVH,subarachnoid
- Focal signs like unequal pupils, hemiparesis- SDH
- Opisthotonic posturing apnea, bradycardia- posterior fossa hemorrhage
- IVH in preterms is usually clinically silent.

3 major steps

- Identify predisposing factors
- Look for early subtle neurological signs.
- Visualise the site,extent and severity by imaging

Imaging modality

- CUS is the first line imaging- subarachnoid ,subdural hemorrahages can be missed
- MRI is more sensitive than CT in detecting haemorrhage-sexcept on the first day
- CT if baby is unstable and when emergencyintervention is required

Management

- Maintain TABC
- Treat seizures with appropriate anticonvulsants
- Look for bleeding or coagulation disorders eg haemophilia if no obvious risk factors
- If a large SDH is suspected LP is contraindicated until a CT scan is done
- PRBC,FFP, vitamin K, cryoprecipitate as necessary
- Most hemorrhages need only conservative management.
- Weekly CUS for detecting hydrocephalus

Indications for neurosurgery

- Signs of brainstem compression- apnea, bradycardia, hypotension
- Acute obstructive hydrocephalus with raised ICT
- Large clot on posterior fossa
- SDH with midline shift

Table for Grading of IVH by imaging

	CUS	CT
Grade 1	GMH with no or minimal IVH (<10% of ventricle)	Isolated GMH
Grade 2	IVH occupies 10-50%	No ventricular dilatation
Grade 3	Occupies >50%, dilated ventricles	Ventricular dilation
Grade 4	Periventricular echodensity	Parenchymal involvement

Neonatal Hypotonia - A Structured Approach

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Introduction

The term "Floppy Infant" is used to describea newborn with poor muscle tone and power(weakness) with increased joint mobility. Hypotonia in a newbornposes a diagnostic challenge, as it is a clinical sign suggestive of both benign and serious conditions. The differential diagnosis for neonatal hypotonia is extensive and a methodical approach helps in localizing the problem to a specific region of the nervous system and formulating a differential diagnosis.

Theunderlying pathology of infantile hypotonia canbe broadly classified into the following categories:

Central (most common)	• Sepsis • Hypoxic ischemic encephalopathy • Intracranial haemorrhage • Cerebral malformations • Chromosomal abnormalities (e.g. Trisomy 21, Prader-Willi syndrome) • Congenital infections (TORCH) • Drug effects (e.g. benzodiazepines, Magnesium toxicity) • Inborn errors of metabolism • Endocrine: hypothyroidism • Benign congenital hypotonia
Spinal cord	• Birth trauma (especially Breech delivery) • Syringomyelia
Anterior Horn Cell	• Spinal Muscular Atrophy • Neurogenic arthrogryposis
Neuromuscular junction	• Myasthenia gravis (transient/ congenital) • Infantile botulism
Peripheral nerves	• Congenital hypomyelinating neuropathy • Hereditary motor and sensory neuropathies (Dejerine-Sottas disease) • Hereditary sensory and autonomic neuropathy • GuillainBarre syndrome (very rare)

Muscle	• Congenital myopathies (e.g. central core disease, Nemaline Rod myopathy, myotubular myopathy, congenital fiber type disproportion and multicore myopathy) • Congenital muscular dystrophies(merosin deficient, Walker - Warburg disease, muscle-eye-brain disease, Fukuyama disease • Muscular dystrophies (inc. congenital myotonic dystrophy)
Metabolic myopathies and multisystem disease	• Disease of Glycogen Metabolism • Acid maltase deficiency • Severe neonatal Phosphofructokinase deficiency • Severe neonatal phosphorylase deficiency • Debrancher deficiency • Peroxisomal disorder • Neonatal adrenoleukodystrophy • Cerebrohepatorenal syndrome(Zellweger's)

Based onclinical criteria hypotonia can be classified in twomajor groups :

1. Central hypotonia
2. Peripheralhypotonia.

Central origin accounts for about 66% to88% of cases, with peripheral origins or unknowncauses accounting for the balance. In additionseveral congenital disorders that are characterizedby hypotonia have both central and peripheralsymptoms, such as metabolic and genetic disorders.

The first goal in diagnosing the source of neonatal hypotonia is to ascertain if it is central (upper motor neuron) or peripheral (lower motor neuron).

A structured approach is necessary in assessing a baby with hypotonia which includes history, initial assessment, examination, and management .

A. History and Initial assessment

- Any significant family history - affected parents or siblings, consanguinity, stillbirths, childhood deaths
- Maternal disease - diabetes, epilepsy, myotonic dystrophy (may not be recognized)
- Pregnancy and delivery history - drug or teratogen exposure
- Decreased fetal movements
- Abnormal presentation
- Polyhydramnios/ oligohydramnios
- Apgar scores
- Resuscitation requirements
- Cord gases
- History since delivery
- Respiratory effort
- Ability to feed
- Level of alertness
- Level of spontaneous activity
- Character of cry

B. Physical Examination

A detailed physical examination should be performed, assessing muscle tone, any asymmetry, the infant's strength, deep tendon reflexes (DTR), and any dysmorphic or unusual features.

	Central	Anterior Horn Cell	Nerve	Neuromuscular Junction	Muscle
Strength	Normal	proximal>distal weakness	distal>proximal weakness	bulbar weakness	proximal>distal weakness
Deep tendon reflexes (DTR)	Normal or increased	decreased/ absent	decreased/ absent	normal	decreased
Abnormal movts	+/- Seizures	+fasciculation	+/- fasciculation		
Others	+/- dysmorphic features	often described as alert			

Here wedescribe a newborn with respiratory distress and hypotonia since the first few hours of life,who was later diagnosed to have nemaline rod myopathy, a rare congenital myopathy

Case report

A term baby boy born to a non-consanguineous G4 P3 mother by normal vaginal delivery. Birth weight was 4165gms. Apgar scores were 7 @ 1mt and 8@ 5mts.

Antenatal scans showed polyhydramnios, bilateral mild renal pelvic dilatation and reduced fetal movements. Baby started grunting soon after birth and gradually worsened, requiring CPAP support. Baby was noted to have poor tone. Full septic screen was done. CXR was taken. On Day 2, NG feeds were initiated but was noticed to have increased oropharyngeal secretions.

On day 5, spoon feeds were tried after weaning off from CPAP, however baby was gagging and secretions increased. Baby continued to have moderate hypotonia

In view of hypotonia, neurology consultation and work up has been done. ECHO, neurosonogram and MRI Brain were normal. CSF studies and blood studies for CPK, Genetic work up and metabolic work up were done which were all normal. EEG was also normal

Baby continued to have swallowing difficulties and hypotonia. EMG of bulbar muscles showed de-nervation - re-nervation injury suggestive of Bulbar palsy

Repeat MRI showed subtle changes in brain stem which were non specific.

Muscle biopsy was taken subsequently which showed thin red rods and variable fibre size suggestive of Nemaline Myopathy

Discussion

The first report of a congenital myopathy was in 1956, when a patient with central core disease (CCD) was described. Since that time, other myopathies have been defined as congenital myopathies

Characteristics of congenital myopathies are as follows:

- Early onset of hypotonia, hyporeflexia
- Frequently have dysmorphicfeatures
- Nonprogressive
- Hereditary
- Unique histo-chemical or ultra-structural features on muscle biopsy

1. Myopathies with protein accumulation
   - Nemaline myopathy
   - Myosin storage myopathy
   - Cap disease
   - Reducing body myopathy
2. Myopathies with cores
   - Central core disease
   - Core-rod myopathy
   - Multiminicore disease
3. Myopathies with central nuclei
   - Myotubular myopathy
   - Centronuclear myopathy
4. Myopathies with fiber size variation
   - Congenital fiber type disproportion

Incidence:

The true incidence of congenital myopathies is unknown.

Fardeau et al documented 180 cases of congenital myopathy over 20 years. The types were as follows:

- Nemaline rod myopathy (20%)
- Central core disease (16%)
- Centronuclear myopathy (14%)
- Minimulticore myopathy (10%)
- Congenital fiber-type disproportion or type 1 fiber predominance (21%)
- Six other miscellaneous congenital myopathies (19%)

Sex:

Both sexes are affected equally in most congenital myopathies since inheritance is usually autosomal recessive or autosomal dominant.

In X-linked forms, boys are affected almost exclusively, although occasional female carriers with clinical manifestations have been described.

Age:

Congenital myopathies usually present in the neonatal period but can also present later in life (even into adulthood).

Clinical presentation:

- Central core disease
  - Early onset with nonprogressive limb weakness, mild facial weakness, and hypotonia
  - History of decreased fetal movement or breech presentation is typical.
  - Skeletal abnormalities may include congenital hip dislocation, kyphoscoliosis, and foot deformities
  - Facial feature include - elongated face, tent-shaped mouth, high-arched palate, and retrognathia are common.

2. Nemaline rod myopathy
   - Early onset . minimally progressive or nonprogressive proximal limb, bulbar, and facial weakness
   - Respiratory insufficiency
   - Skeletal deformities include arthrogryposis , limb contractures, kyphoscoliosis, pectusexcavatum, and rigid spine
   - Cardiomyopathy
   - CNS disease is rare, but seizures have been reported in severe cases

3. Centronuclear/ Myotubular myopathy

Three different presentations have been described

A. Autosomal dominant form:
   - Hypotonia at birth and poor suck
   - Facial weakness, high-arched palate
   - Ptosis ; Ophthalmoplegia
   - Joint hyperlaxity, and contractures
   - Weakness is distal more than proximal

B. Severe X-linked form:
   - Decreased fetal movements and polyhydramnios in utero
   - Severe weakness and hypotonia with feeding difficulty and respiratory distress
   - Bilateral ptosis facial weakness and ophthalmoplegia
   - Skeletal abnormalities include pectuscarinatum, micrognathia, knee and hip contractures

C. Autosomal recessive form:
   - Slowly progressive course
   - Reduced fetal movements and oligohydramnios
   - Hypotonia with proximal weakness
   - Facial weakness, ptosis, and ophthalmoplegia
   - Dilated cardiomyopathy
   - Mental retardation

4. Multiminicore disease
   - Nonprogressive or minimally progressive
   - Proximal and axial weakness and hypotonia
   - Facial and bulbar weakness

- Progressive respiratory insufficiency
- In non - classical forms , ophthalmoplegia and skeletal abnormalities have been reported

5. Congenital fiber-type disproportion
   - Hypotonia and diffuse weakness (Type 1 muscle fibers affected)
   - Facial, bulbar, and respiratory weakness
   - Short stature , low body weight
   - Multiple joint contractures and scoliosis

A suggested protocol for differential diagnosis of Congenital Hypotonia (De Vivo et al)

Signs and symptoms	Type	I Level	II Level	Diagnosis
Hypotonia Weakness Hypo-areflexia Respiratory failure Poor cry / suck Family/Maternal history suggestive	**Peripheral Hypotonia**	Muscle enzyme (CK, LDH)	EMG, muscle biopsy	If suggestive, search for mutations - Hereditary neuropathies (SMA I) - Congenital muscular dystrophies -Congenital myopathies
Hypotonia Sensory impairment Seizures, Hypo-hyperreflexia Personal and pregnancy history suggestive	**Central Hypotonia**	Screening for sepsis , ammonia, EEG and/or aEEG	Neuroimaging (NSG , CT and/or MRI)	Congenital trauma, HIE, HIC, infections, brain malformations
Mixed symptoms	**Mixed Hypotonia**	Multidisciplinary approach (Geneticists, Neurologists, experts on metabolism)	Karyotype, molecular analysis, FISH studies	Chromosomal rearrangements - trisomy: 13, 18, 21 - Subtelomeric deletions (+ tests of methylations) Prader-Willi's syndrome -Metabolic and storage diseases

Nemaline Rod Myopathy

Nemaline myopathy is defined by muscle weakness and the presence of fine, thread-like or rod-like structures called "nemaline bodies" in muscle biopsies.

"Nema" is derived from Greek and means "thread-like."

Nemaline bodies consist of accumulations of muscle proteins.

The major clinical features are muscle weakness, hypotonia and reduced or absent reflexes

Muscle weakness is usually most severe in muscles of the face, neck and proximal muscles.

Six different clinical subtypes of nemaline myopathy have been identified based on disease severity and age of onset, ranging from a severe congenital-onset (at birth) form that is usually lethal in the first few months of life, through to less severe forms with onset in childhood or adulthood

1. Typical Congenital Nemaline Myopathy
2. Severe Congenital (Neonatal) Nemaline Myopathy
3. Intermediate Congenital Nemaline Myopathy
4. Childhood-Onset Nemaline Myopathy
5. Adult-Onset Nemaline Myopathy
6. Amish Nemaline Myopathy

Nemaline myopathy can be inherited as an autosomal recessive or dominant trait.

At least 50% of cases of nemaline myopathy follow autosomal recessive inheritance, and the remainder are inherited in an autosomal dominant manner or are sporadic

Mutations in the ACTA1 gene have been found to cause approximately 15-25 percent of nemaline myopathy. ACTA1 mutations may cause the severe, intermediate or typical congenital forms of nemaline myopathy.

Mutations in the NEB gene have been identified as a cause of about 50% of nemaline myopathy.

Most individuals with a NEB mutation have the typical congenital form.

Mutations of the NEB gene are inherited as an autosomal recessive trait.

Diagnosis of nemaline myopathy is suspected based upon a thorough clinical evaluation, a detailed patient and family history and identification of characteristic findings.

Diagnosis is confirmed by the presence of thread- or rod-like structures (nemaline bodies) on muscle biopsy when stained with Gomoritrichrome.

No specific treatment is available for any of the congenital myopathies, but aggressive supportive care is essential to preserve muscle activity, to allow for maximal functional ability, and to prolong life expectancy.

Genetic counselling is needed for the affected families

Suggested readings:

Fenichel GM. Neonatal Neurology 3rd edition. Churchill Livingston Inc. 1990

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Approach to common neonatal surgical emergencies

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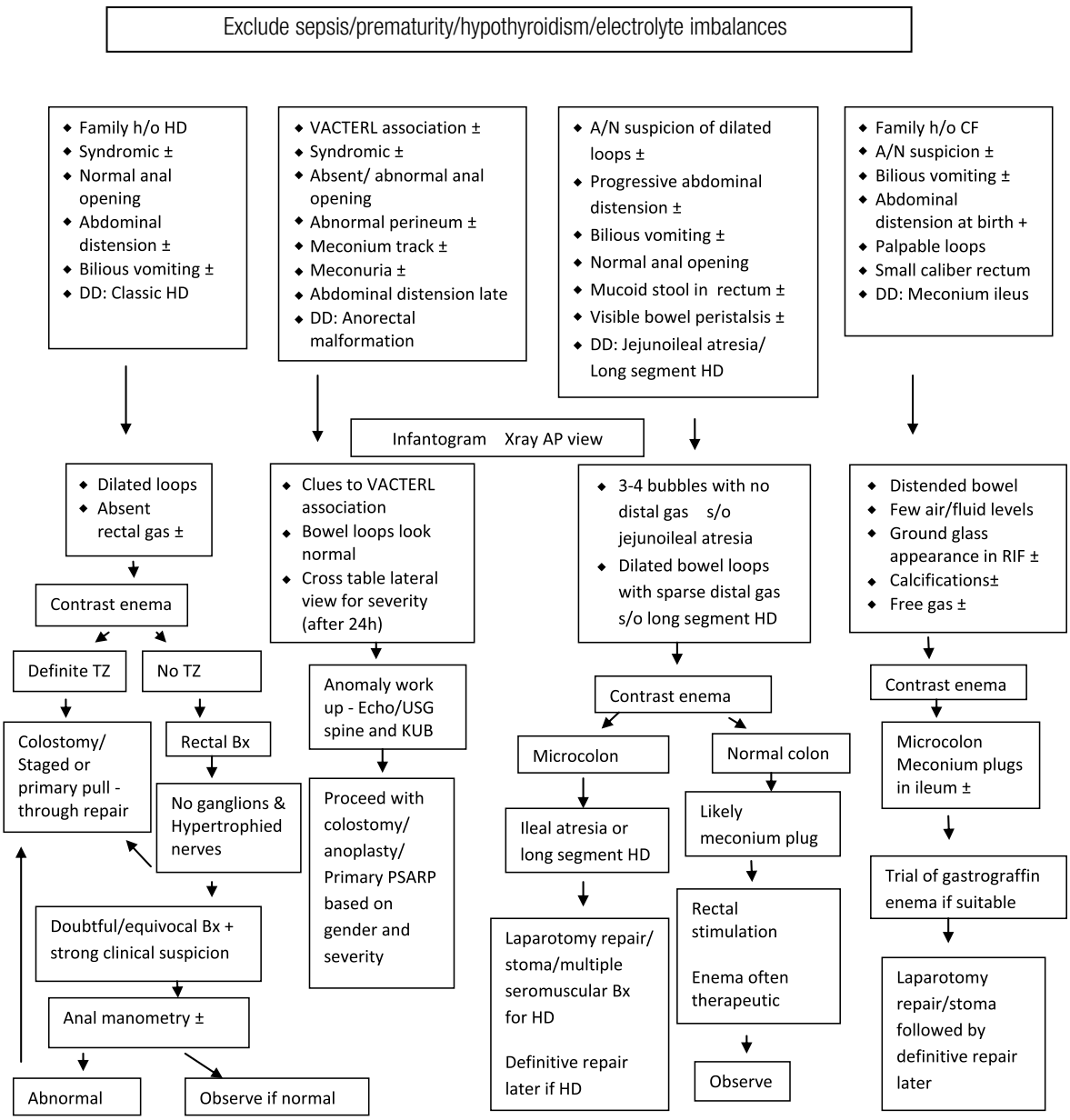
General guidelines:

1. Check antenatal pointers to diagnoses such as family history, ultrasound findings
2. Look for anomalies that can present as associations
3. Narrow down on surgical possibilities before special investigations
4. A simple infantogram (Xray - neck to knee AP view) can provide a lot of information
5. Almost always the etiologies are congenital
6. Cross consult when in doubt; two minds work better than one!
7. Not all surgical emergencies need immediate operative interventions
8. Serial examination and a period of observation are required in many cases for decision making

Common clinical scenarios:

- Failure to pass/delayed passage of meconium
- Bilious vomiting
- Respiratory distress
- Poor urinary stream/straining during voids
- Discoloration/Swelling inguino-scrotal region
- Frothing at the mouth or choking on initial feeds
- Prolapse of bowel loops outside abdomen at birth
- Non bilious vomiting

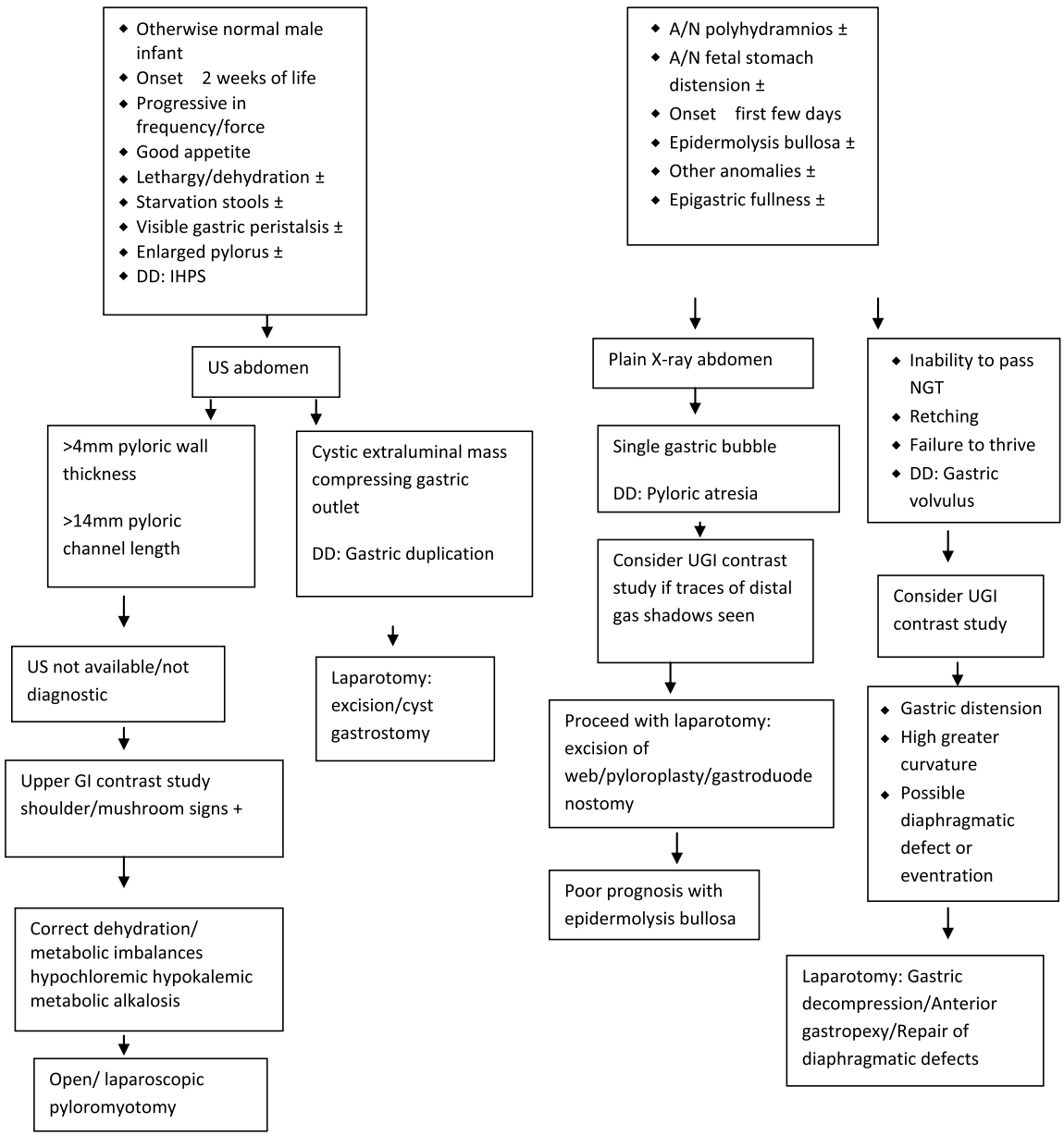
Failure to pass / delayed passage of meconium (>48 h)



Abbreviations: HD - Hirschsprung's disease; A/N - antenatal; CF - cystic fibrosis; TZ - transition zone; DD - differential diagnosis; Bx - biopsy, RIF - right iliac fossa, PSARP - posterior sagittal anorectoplasty

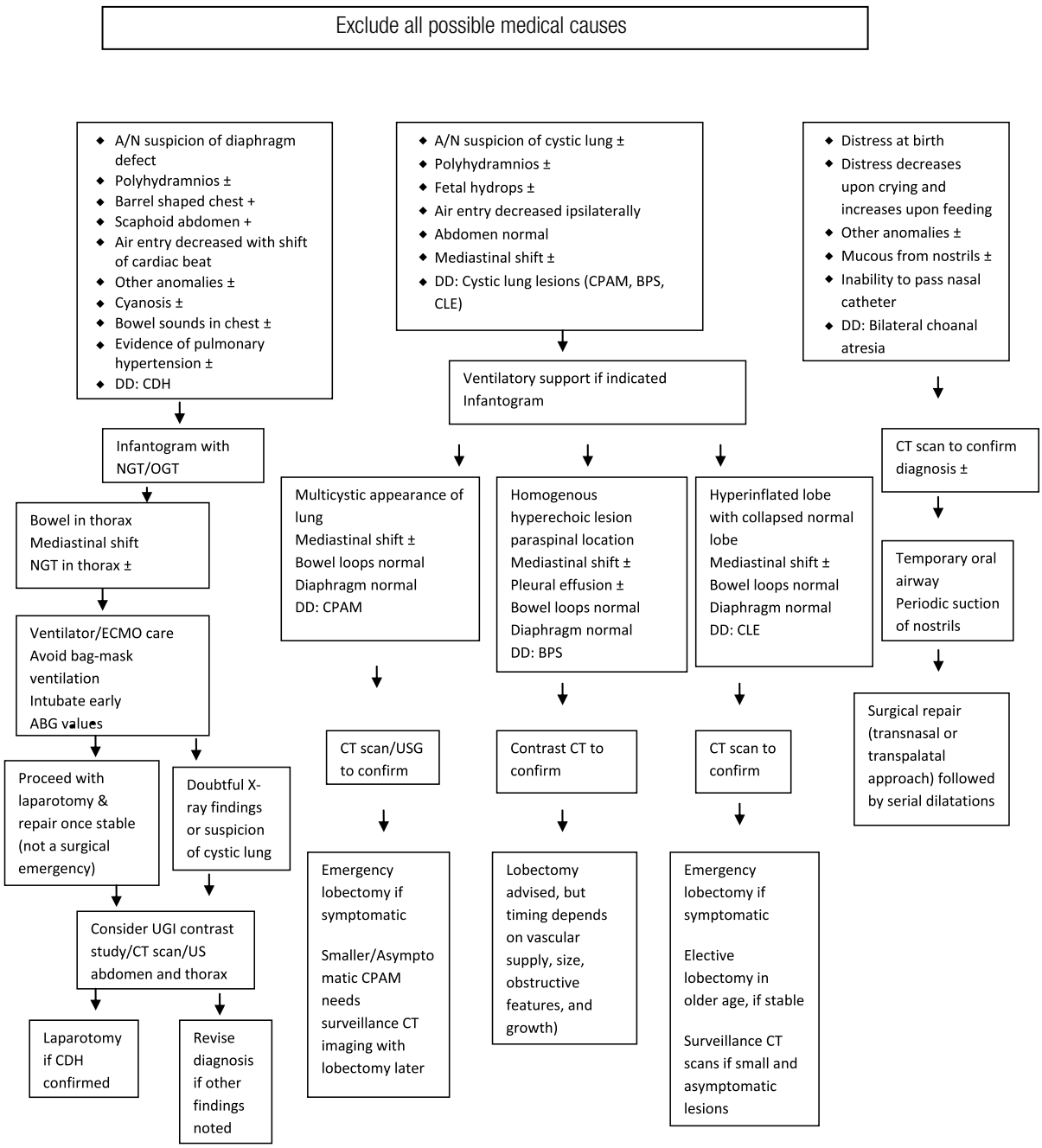
Non bilious projectile vomiting

Exclude gastroenteritis/raised ICP/UTI/metabolic disorders/CAH/food allergy

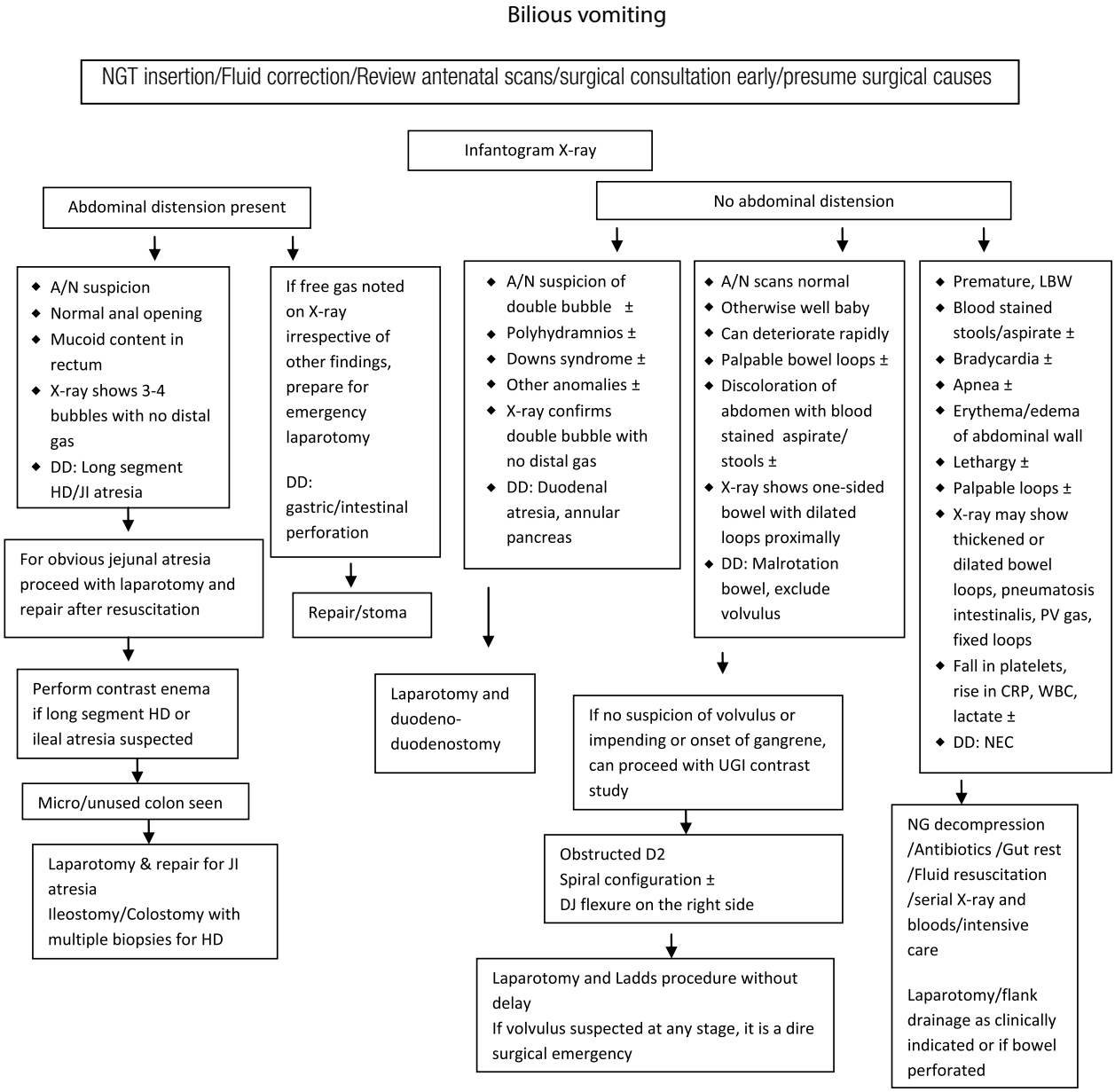


Abbreviations: ICP - Intracranial pressure, UTI- Urinary Tract infection, CAH- Congenital adrenal hyperplasia, IHPS- infantile hypertrophic pyloric stenosis, A/N- antenatal, US- Ultrasound, DD- differential diagnosis, UGI - upper GI

Respiratory distress in newborn - surgical

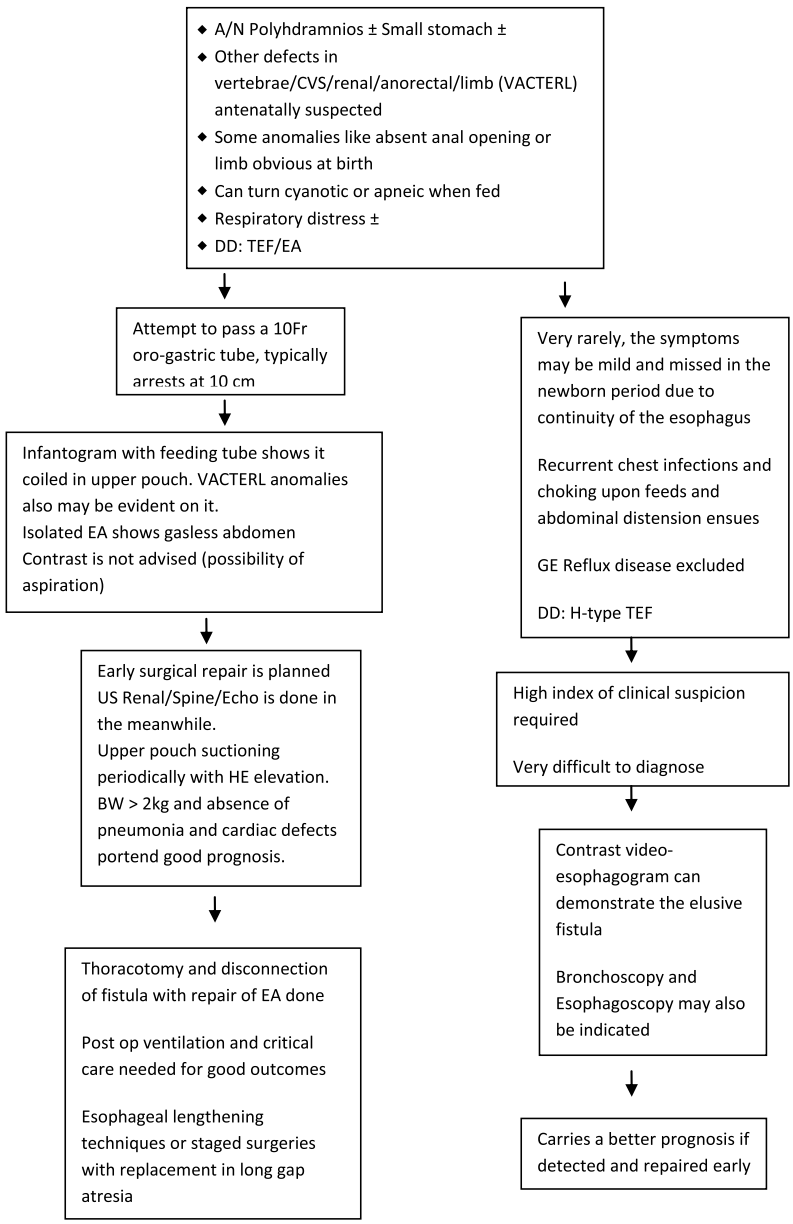


Abbreviations: CPAM- Congenital Pulmonary Airway malformation, BPS- Bronchopulmonary sequestration, CLE- Congenital lobar emphysema, A/N- antenatal, US- Ultrasound, DD- differential diagnosis, CDH- Congenital diaphragmatic hernia



Abbreviations: A/N- antenatal, US- Ultrasound, DD- differential diagnosis, HD - Hirschsprung's disease, JI - jejunoileal, D2 - second part of duodenum, PV - portal vein, CRP - C reactive peptide, NEC - nectrotising enterocolitis, WBC- White blood cells, DJ- duodenojejunal, LBW - Low birth weight babies

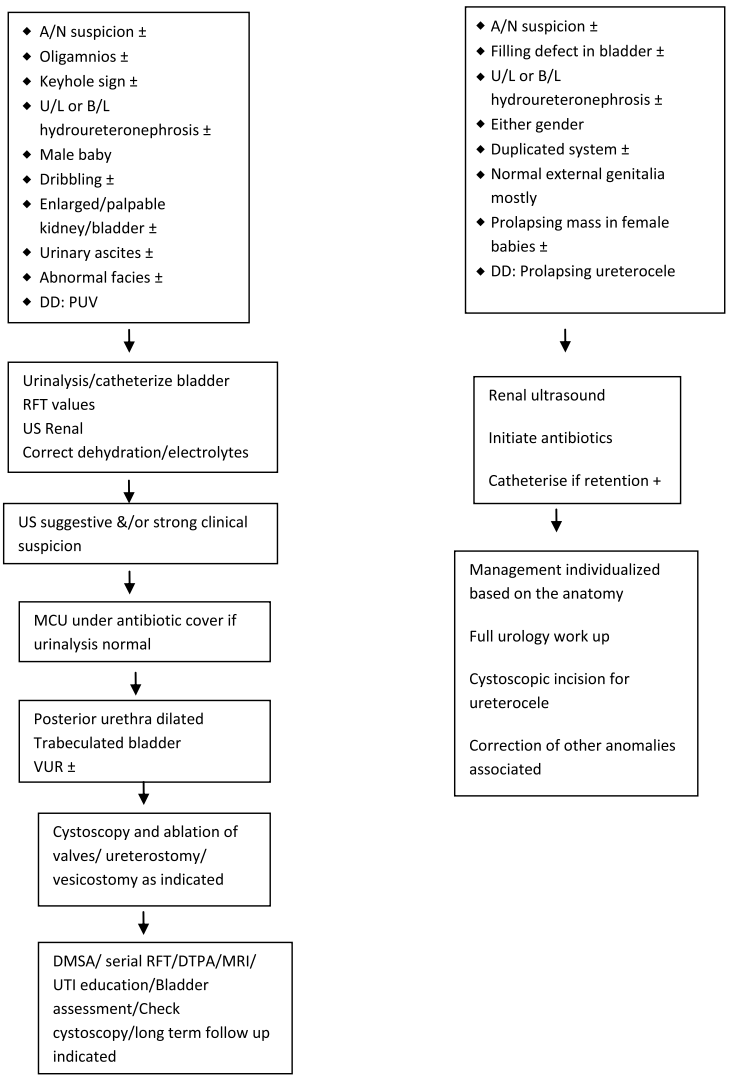
Frothing at the mouth or choking during initial feeds



Abbreviations: A/N- antenatal, DD - differential diagnosis, CVS- Cardiovascular system, VACTERL- Vertbral/Anorectal/Cardiac/Tracheo-esophageal/Renal/Limb, EA- Esophageal atresia, TEF- Tracheo-esophageal fistula, BW - Birth weight, US - Ultrasound

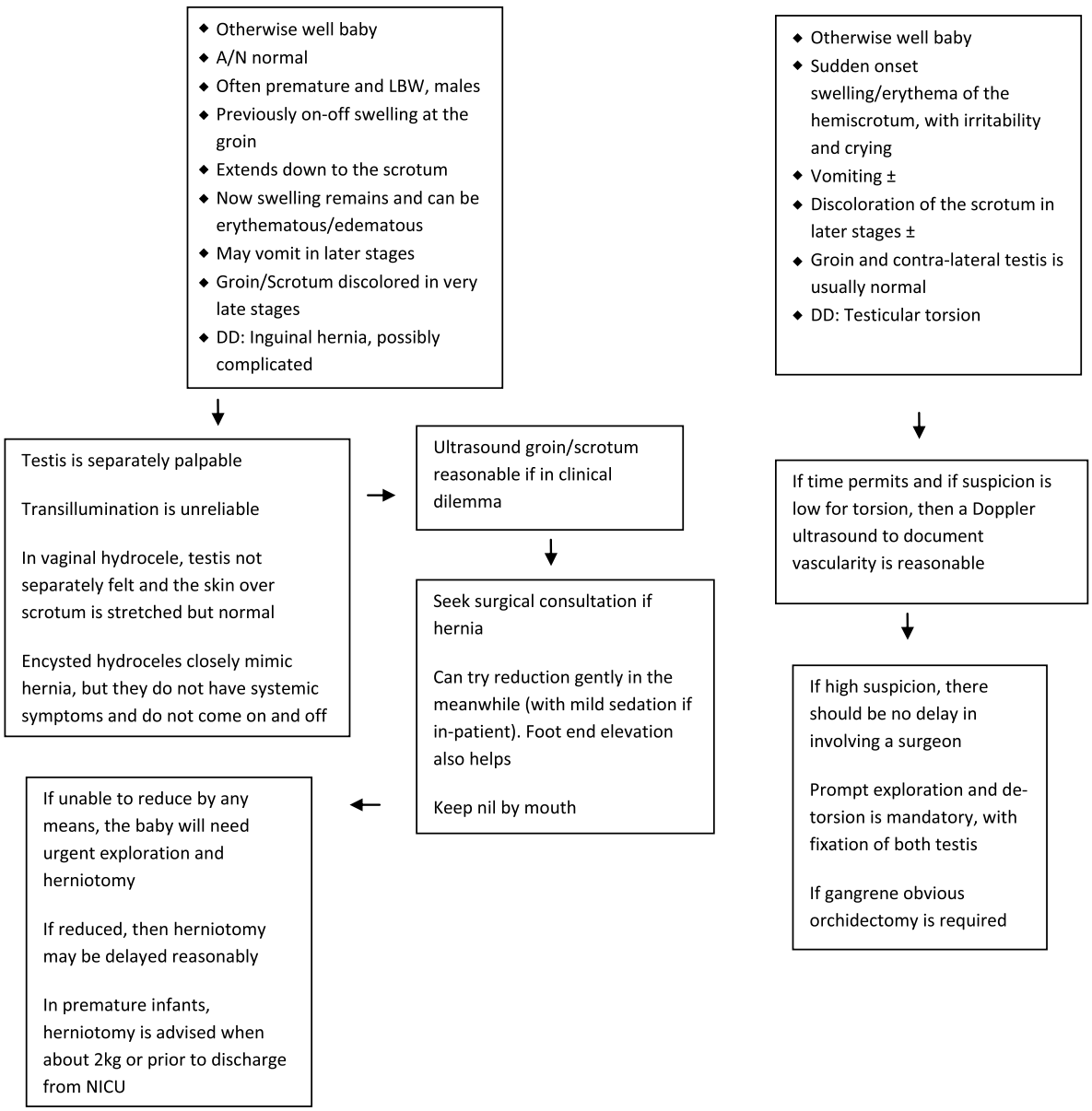
Poor urinary stream/ Straining during voids

Exclude abnormalities of the external genitalia/spine



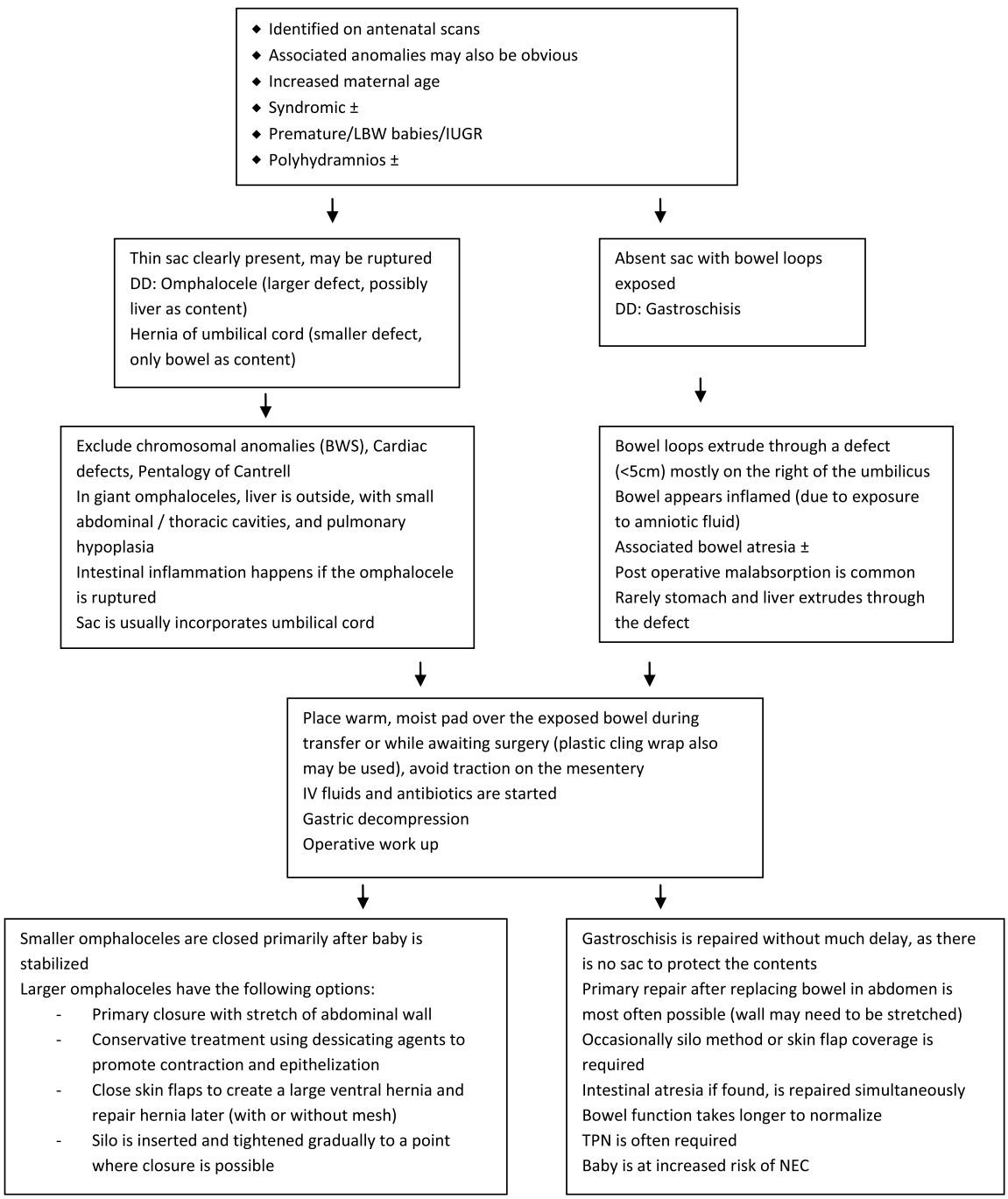
Abbreviations: A/N - antenatal, DD - differential diagnoses, U/L - unilateral, B/L - bilateral, RFT - renal function tests, MCU - Micturating cystourethrogram, VUR - Vesicoureteral reflux, DMSA/DTPA - isotope scans, UTI - urinary tract infection, MRI- magnetic resonance scan

Discoloration/Swelling at the inguino-scrotal region



Abbreviations: A/N - antenatal, DD - differential diagnoses, NICU - newborn intensive care unit

Discoloration/Swelling at the inguino-scrotal region



Abbreviations: DD - differential diagnoses, LBW - low birth weight, IV - intravenous, IUGR - intrauterine growth retardation, TPN - total parenteral nutrition, NEC - necrotizing enterocolitis

Respiratory Distress in the Newborn -An Overview

Dr. PMC Nair

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Respiratory distress in the newborn is the second commonest cause of emergency admissions in the NICU.

Symptoms include tachypnea (respiratory rate >60 /minute), apnea or dyspnea, grunting, inspiratory stridor, nasal flaring, suprasternal, intercostal, subcostal retractions, cyanosis and poor feeding.

Most cases are caused by transient tachypnea of the newborn (TTN), Respiratory distress syndrome (RDS) or Meconium aspiration syndrome (MAS). (Table1).

Clues to diagnosis: (even without a stethoscope!!)

1. If baby has respiratory distress with suprasternal retractions, it has to be an upper airway obstruction.
2. If baby has respiratory distress with intercostal and subcostal retractions, it has to be a lung parenchymal disorder and if associated with whimpering or moaning there may be associated pneumonia, shock or CNS involvement.
3. If it is a preterm with triad of tachypnea, intercostal retractions and expiratory grunt, it is RDS (HMD)
4. If H/o LSCS, late preterm or early term baby with only soft tachypnea and X-ray shows fluid in theinterlobar fissure and perihilar shadows, it is retained lung fluid or TTN
5. If respiratory distress and there is history of maternal GBS, Chorio-amnionitis, maternal fever, PROM etc think of Pneumonia.
6. History of MSAF, post term , fetal distress or perinatal depression and hyperinflated chest, think of MAS
7. If history of polyhydramnios in the mother and baby has frothing in the mouth, and respiratory distress think of esophageal atresia with tracheo-esophageal fistula.
8. If respiratory distress with dextrocardia, and decreased air entry on the left side of chest with gurgling sounds (borborgymi) and flat or scaphoid abdomen, think of Congenital diaphragmatic hernia
9. If baby has cyanosis and mild tachypnea, think of cardiac cause of respiratory distress
10. If there is history of oligohydramnios, renal dysplasia or agenesis, neuromuscular disorder (bell-shaped chest),or diaphragmatic hernia and respiratory distress, think of possibility of pulmonary hypoplasia

Table: Causes of Respiratory Distress in the Newborn

Pulmonary /non-pulmonary causes

Pulmonary: Upper airway/lower airway

Airway: Nasal obstruction, Choanal atresia, Pierre Robin sequence, macroglossia, laryngeal web or cyst, laryngomalacia, trachea-esophageal fistula, vascular rings, external compression from a neck mass, vocal cord paralysis, hemangiomas , subglottic stenosis

Pulmonary: RDS, TTN, MAS, Pneumonia, Pneumothorax, PPHN, pleural effusion(congenital chylothorax), pulmonary hemorrhage, congenital cystic adenomatoid malformation (congenital pulmonary airway malformation), pulmonary hypoplasia, congenital lobar emphysema, pulmonary alveolar proteinosis, alveolar capillary dysplasia, surfactant protein deficiency

Cardiovascular: Critical Congenital heart Disease, cardiac failure, neonatal cardiomyopathy, pericardial effusion, cardiac tamponade.

Thoracic: Chest wall deformities, Congenital diaphragmatic henia, paralysis, asphyxiating thoracic dystrophy, Pneumomediastinum

Neuromuscular: CNS injury (birth trauma/ hemorrhage), HIE, cerebral malformations, chromosomal abnormalities, torch infections, meningitis, seizure disorder, arthrogryposis, neonatal myasthenia gravis, spinal muscular atrophy, congenital myopathies,maternal or neonatal sedation.

General: Metabolic disturbances, like hypoglycemia, hypo or hypernatremia, hypermagnesemia, IEM, metabolic acidosis, anemia, polycythemia, sepsis.

Conclusion:

Failure to readily recognize symptoms and treat the underlying cause of respiratory distress can lead to short- and long-term complications and related mortality in at-risk infants.

Five ventilator strategies for neonatal intensivists in the management of respiratory Distress

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Respiratory management of an acutely ill neonate has evolved continuously with newer insights in the last two decades.The following is a snapshot summary of the current trend and newer strategies in the ventilator management of a neonate with respiratory distress:

1. Non Invasive Ventilation: The most important strategy for providing respiratory support to a spontaneously breathing neonate is by maintaining functional (FRC) through positive end expiratory pressure (PEEP). This can be done very effectively by CPAP (continuous positive airway pressure) and NIPPV (nasal intermittent positive pressure ventilation) in all neonates. HHHFNC (heated humidified high flow nasal canula) is another baby friendly alternative that can be tried in more stable neonates above 28 weeks gestation.
2. Gentle ventilation: Accepting higher pCO2 levels (45-55) and lower pH levels (7.25 - 7.35) in the blood gas, optimizing FiO2 requirements to target an O2 saturation between 90 and 94 (paO2: 60 -80) and choosing baby friendly ventilation modes like SIPPV over SIMV, use of ventilator modes that cycle into expiratory phase by flow triggering rather than time triggering like PSV whenever possible are all accepted strategies to prevent lung injuries. Real time monitoring to prevent hyperventilation by active use of pulmonary graphics is another strategy for faster weaning of neonates off ventilator.
3. Volume Targeted Ventilation: Conventional neonatal ventilation modes like SIMV and SIPPV that limit the peak inspiratory pressure and hence limit the barotrauma does not prevent volutrauma caused by the excessive tidal volumes as the lung condition improves on a ventilator. Volutrauma which is more damaging in neonates can be more effectively prevented by the use of a hybrid mode (Volume Guarantee mode or targeted tidal volume mode along with SIPPV or PSV) that limits peak volume rather than peak pressure[1,2,3].For volume targeted ventilation to be successful, we need to remember that the dead space ventilation is more in smaller babies and hence we need to target larger tidal volumes (5-6 ml/kg) in preterm neonates as compared to smaller volumes (4-5 ml/kg) in late preterm and term neonates.
4. Elective use of HFO?: Use of smaller tidal volumes and the safer use of higher mean airway pressure are the potential advantages of HFOV over conventional mechanical ventilation. However older studies that have looked at the superiority of HFO over conventional ventilation did not find any difference between the two probably because of the late conversion of sicker babies from conventional ventilation to HFO.Newer insights (Courtney et al, Johnson et al, etc) highlighting the importance of early conversion are emerging since 2002. According to the results of the latest Cochrane review[4], the use of elective HFOV compared with conventional mechanical ventilation results in a minor reduction in the risk of chronic lung disease. The evidence is however weakened by the inconsistency of this effect across trials.
5. HFO with VG?: A newer concept of volume-targeted (VG) ventilation during HFOV has been introduced in some new generation neonatal HFOV devices recently. Volume-targeted ventilation is known to improve neonatal prognosis in preterm infants when added to conventional ventilation. However, volume-targeted ventilation combined with high frequency ventilation has not been adequately evaluated. In a prospective, randomized, short term crossover clinical study that compared HFOV with and without VG in neonates with acute RDS, because of the lower VThf fluctuation and lower incidences of out-of-target PCO2 levels, HFOV combined withVG was suggested to be better for preterm infants[5]. Another pilot study by Martin Kesler, et al suggests that VG combined with HFOV attenuates fluctuation of SpO2 and CO2 clearance, which may prevent hypoxemia and hypocapnia[6]. (PS: Our unit experience: In our limited experience at Jubilee, we found the use of HFO with VG highly encouraging. A poster of our pilot study was presented by us in this year's state conference of NNF Kerala: CalNeocon 2017).

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6. Enomoto, Kesler, et al. A Pilot Study of Volume Guarantee with HFOV, American Journal of Perinatology · May 2016 DOI: 10.1055/s-0036-1584141

Approach to a Septic Neonate

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Emergency Signs in TRIAGE/ER

The Septic neonate presents with danger signs

- Hypotherma
- Gasping respiration
- Severe Respiratory Distress
- Central cyanosis
- Shock
- Bulging AF/Coma/lethargy
- Bleeding Tendency

Admit in NICU

Assessment

•	S	Sensorium	AF, Active / Lethargic / Tone Difference
•	T	Temperature	Hypothermia <36.5 C
•	O	Oxygen Saturation	with Oxygen / without Oxygen
•	P	Perfusion/ Shock	CFT >3s
			mottling
			acrocyanosis
			NIBP
•	S	Sugars	Hyperglycemia
			hypoglycemia
•	S	Skin	Mottling / Deep Jaundice
			Bleeding tendency
•	S	Other Features	Hepatosplenomegaly
			Respiratory Distress

History

< 72 hrs	72 hrs
EONS	LONS
• Prematurity / LBW	• VLBW Baby
• PROM > 18 hrs	• Prolonged ICU stay
• Maternal Pyrexia	• Exposure to antibiotics
• UTI / Chorioamnionitis	• Central venous catheters
• Prolonged labour	• TPN / Intubation
• Instrumental Delivery	• Prolonged Ventilation
• Perinatal hypoxia	• Contact history of fever at home
• Apgar <4	• NEC

Clinical Examination for Subtle / Non Specific signs

- Hypothermia / Fever
- Letharagy/ Poor suck
- Poor Perfusion , prolonged CFT
- HRC(Heart Rate Changes) ,Apnea
- Hypo / hyperglycemia
- Metabolic Acidosis

Specific features related to various systems:

Central nervous system (CNS): Bulging anterior fontanelle, vacant stare, high-pitched cry, excess irritability, stupor/ coma, seizures, neck retraction (clinical suspicion of meningitis)

Cardiac: Hypotension, poor perfusion, shock, Loss of heart rate characterestics (HRC)

Gastrointestinal: Feed intolerance, vomiting, diarrhea, abdominal distension, paralytic ileus, necrotizing enterocolitis (NEC)

Hepatic: Hepatomegaly, direct hyperbilirubinemia (especially with urinary tract infections)

Renal: Acute renal failure

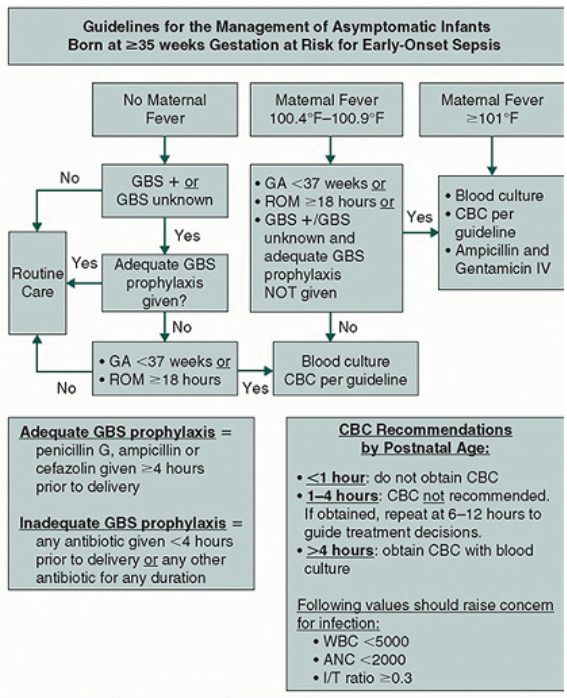
Hematological: Bleeding, petechiae, purpura

Skin changes: Multiple pustules, abscess, sclerema, mottling, umbilical redness and discharge.

Epidemiology: Indian data

The incidence of neonatal sepsis according to the data from National Neonatal Perinatal Database (NNPD, 2002-03) is 30 per 1000 live births. One of the commonest causes of neonatal mortality 19% of all neonatal deaths (3).

Among intramural births, Klebsiella pneumoniae was the most frequently isolated pathogen (32.5%), followed by Staphylococcus aureus (13.6%). Among extramural neonates (referred from community / other hospitals), Klebsiella pneumoniae was again the commonest organism (27%), followed by Staphylococcus aureus (15%) and Pseudomonas (13%) (3).



LAB

Rapid Diagnostic Tests

C-reactive protein : Among the acute phase reactants, CRP, produced in the liver, is a frequently used laboratory test for the diagnosis of neonatal sepsis. This biomarker has a half-life of 24-48 hours. Importantly, CRP takes 10-12 hours to respond after an infection, making it an unreliable marker of the initial stages of an acute infection. Given its response rate, serial CRP levels in combination with the absolute and complete WBC and I/T ratio has been widely used as a negative predictor of sepsis 24-48 hours after the onset of symptoms.

- CRP : better after 6 hrs of birth in EOS
  at least 2 serial measurements

* ANC/m3 : > 1750 Significant, <1000 High positive value

WHITE BLOOD CELL COUNT (NEUTROPHIL INDICES)

- Absolute neutrophil count,absolute band count, and immature to total neutrophil (I/T)ratio-to identifyinfected infants.

- Neutropenia better marker , better specificity
- Few conditions besides sepsis(maternal pregnancy-induced hypertension, asphyxia, and hemolytic disease) depress the neutrophil count of neonates.
- In late preterm and term infants, the definition for neutropenia most commonly used is that suggested by Manroe et al(<1800/mm3 at birth and <7800/mm3at 12-14 hours of age).
- TLC Leukopenia more suggestive .Use charts
- < 4000/m and >15000 suggestive.
- 1/T ratio 0 .3 more suggestive Immature/ total WBC (0.27 is the 90th centile - The I/T ratio best sensitivity of any of the neutrophil indices.
- The I/T ratio is<0.22 in 96%of healthy preterm infants age.
- Maximum normal values for the I/T ratio occur at birth (0.16)
- In healthy term infants, the 90th percentile for the I/T ratio is 0.27.59
- Micro ESR Age in days + 6 (>15mm)

2 Parameters to be considered to rule out sepsis

Platelet Counts

Despite the frequency of low plateletcounts in infected infants, they are nonspecific, insensitive, and late indicator of sepsis.

Procalcitonin : Appears 2 hrs after invasion and peaks by 4 - 5 hrs. Value falls down early with subsidence of infection

Blood Culture : Gold STD

Minimum 1 ml Blood to moculated. Tken 24 - 48 hr.

Newer methods BACTEC METHOD. Useful while on antibiotics.

Early detection of Signals

Urine Culture : SPC Gold Std

Xray Chest congenital pneumonia

CSF Study :

- Symptomatic cases of EOS not responding to Antibiotics
- Symptomatic LOS
- Not in ASYMPTOMATIC CASES of suspected EOS.
- HMD where antibiotics started for preterms
- UNSTABLE vitals.

Analyse

- Cells
- Sugar
- Protein

CULTURE AND SENSITIVITY

WITH SUSPECTED SEPSIS	WITH PROVEN SEPSIS
PRETERM	TERM
WBC > 25 AND	WBC > 10 OR
Protein> 170 OR	Glucose < 25 OR
WBC > 100 OR	Protein > 170
Glucose < 25 mg	WBC > 8 or
TERM	
WBC > 21 or Glucose < 20 mg	Glucose < 20 mg/dl OR Protein > 12

Supportive Care

- Oxygen Saturation in normal range by O2/CPAP/ mechanical ventilation
- Nurse in a thermo / neutral environment to avoid hypothermia / hypertherma
- Anemia, thrombocylopenia, DIC are treated with appropriate transfansfusions
- Hypoglycerma to be corrected.>60 mg/dl

Shock

- I/V Bolus NS 10 - 20 ml /Kg over 20 - 30 minutes
- Correct hypoglycemia
- Inotropes, Dopamine 5 - 15microgm/kg/minute

Hydrocortisone1-2mg /kg/q8H if response to Dopamine is not adequate

Urine analysis

When there is FTT, fever, vomiting, diarhoea, jaundice, irritability, VLBW infant known urinary tract anomalies, visibly turbid urine to be investigated.

SPA Sample to be used always for urine c/s or by a fresh catheter

Antibiotic Treatment

As per the antiobiogram of the nursery

I line :- Ampicillin and Amkacin. Some centres start on ciproflox-acin and amkacin

II line :- Piptaz and Amikacin/vancomycin

III line :- Meropenem and vancomycin

To replace with Cefotaxim and amikacin for treatment of meningitis and meropenem and amikacin in resistant meningitis. In NEC a gram positive, gram negative and anaerobic cover with metronidazole or meropenem is advised.

Imp: "Emperical use of 3rd generation Cephalosporin to be avoided especially cefotaxim.

Fungal Infection

- Risk factors
- Prematurity
- VLBW
- Prolonged NICU stay
- Prolonged Central venous catheters
- Use of broad spectrum antibiotics
- Cephalosporins and carbapenams
- Hydrocortisone, PPI, TPN, prolonged intubation

In nurseries with high prevalence of candidiasis, prophylaxis with antifungals in VLBW babies Fluconazole 3 - 6 mg/kg/day twice a week oral / IV till 28 days of life in VLBW babies

Proven cases - Injection fluconazole 5 - 6 mg/kg/ IV OD

Complicated cases (meningitis, endoc)

Inj liposomal Ampho B 5 - 7 mg / kg / IV is given

After culture report is available

- Sensitive antibiotic with a narrow spectrum is used
- Single sensitive antibioticcan be used except in pseudomonas infection
- If the neonate deteriorates in sensitive antibiotics can change

Table 49.1. Suggested Antibiotic Regimens for Sepsis and Meningitis

Organism	Antibiotic	Bacteremia	Meningitis
GBS	Ampicillin or penicillin G	10 days	14-21 days
Escherichia coli	Cefotaxime or ampicillin and gentamicin	10-14 days	21 days
CONS	Vancomycin	7 days	14 days
Klebsiella, Serratia[3]	Cefotaxime or meropenem and gentamicin	10-14 days	21 days
Enterobacter, Citrobacter[3]	Cefepime or meropenem and gentamicin	10-14 days	21 days
Enterococcus[3]	Ampicillin or vancomycin and gentamicin	10 days	21 days
Listeria	Ampicillin and gentamicin	10-14 days	14-21 days
Pseudomonas	Ceftazidime or piperacillin/tazobactam and gentamicin or tobramycin	14 days	21 days
Staphylococcus aureus[23]	Nafcillin	10-14 days	21 days
MRSA	Vancomycin	10-14 days	21 days

Antibiotics

If no senstive antibiotic reported, moderately senstive dose at highest dose is used.

Duration

- Blood culture postive - 14 days
- If culture negative at 48 hrs and asymptomatic baby without risk - STOP
- Suspected EOS/LOS and completely asymptomatic -STOP

LOS / EOS improves not completely

CRP +ve - antibiotics : 7d

CRP ve - antibiotics *5 days

Meningitis *21 days

- Monitor OTC weekly
- I/o chart monitoring
- Hearing at 4 - 8 weeks
- NSG at 1st week
- Repeat LP not a routine
- if not improving
- NSG abnormaL
- SEPSIS MIMICS

  EONS:RDS.ASPHYXIA.

  LONS :IEM ,CCHD DUCT DEPENDENT LESIONS

Table 2. Effective current measures and new approaches to prevent, diagnose, and treat neonatal sepsis.

Category	Measure	Item
Prevention	Current measures	Improved maternal health and nutrition
		Clean delivery practices and handwashing
		Risk-based intrapartum antibiotic prophylaxis
		Hand washing from health care providers
		Promotion of early initiation of exclusive breastfeeding
	New approaches	Maternal *S. agalactiae* and *S. pneumoniae* immunization
		Maternal vaginal chlorhexidine other antisepsis preparations
		Neonatal protective and antisepsis skin preparations
		Neonatal vitamin A supplementation
		Recombinant active antimicrobial proteins
		Toll like receptor agonists
Diagnosis	Current measures	Blood culture
		Antigen detection
		Blood neutrophil count and differential
		C-reactive protein
	New approaches	Proteomic, amniotic fluid analysis
		Improved clinical syndrome identification
		Real-time polymerase chain reaction
		Interleukin inflammatory indices
		Microfluidic macrotechnologies
		"Lab on a chip" point of care devices
Treatment	Current measures	Parenteral antibiotics (penicillin/ampicillin and gentamicin or third generation cephalosporins) for 10-14 d
	New approaches	Shorter courses of antibiotic therapy
		Better-absorbed oral antibiotics especially second generation cephalosporins and ciprofloxacin

RECENT ADVANCES

Developing biomarkers

Acute phase proteins and other proteins

Serum amyloid A : Serum amyloid A (SAA) is an apolipoprotein pro-duced in the liver and an early acute phase reactant that has been studied, though not extensively, in neonates. SAA is derived from a variety of other tissues such as endothelial cells, monocytes, and smooth muscle cells and regulated by cytokines IL1 and IL6 as well as TNF?.

Lipopolysaccharide-binding protein

Lipopolysaccharide-binding protein (LPB), primarily produced by hepatocytes but also by epithelial and muscle cells, is a soluble pat-term-recognition molecule important for interaction with endotoxin of gram-negative bacterial infections. LPB recognizes microbial-associated molecular patterns of bacteria to transport endotoxin to CD14 immune effector cells in response to infections. Binding to the lipopolysaccharide component of the bacteria .

Cytokines and chemokines

Functionally classified, proinflammatory cytokines include cytokines such as interferon-gamma, TNF?, inducible protein -10 (IP-10), and IL-2, IL-6, IL-8, IL-12, and IL-17. Multiple function inflammatory cytokines include cytokines such as IL-1?, monocyte chemoattractant protein (MCP-1), and soluble CD40 ligand (sCD40L) and growth factors, IL-3, granulocyte-colony stimulating factors and their secondary mediators, nitric oxide, thromboxanes, leukotrienes, platelet-activating factor, prostaglandins, and complements. These multiple function secondary mediators cause activation of the coagulation cascade, the complement cascade, as well as participate in the production of prostaglandins, leukotrienes, proteases, and oxidants.

Cell-surface antigens

Circulating inflammatory cells such as neutrophils, lymphocytes, monocytes, and natural killer cells express cell surface antigens, after activation by microbial products, that can be detected by flow cy-tometric technology. Several cell surface antigens such as CD11b, CD 14, CD64, CD32, CD16, CD69, and sCD163 have been identified to be promising in the detection of congenital sepsis, as well as early and late onset neonatal sepsis.

Antigen detection techniques allow rapid detection and identification of microorganisms without culturing.

The most commonly used commercially available test is the latex ag-glutination assay, which is based on specific agglutination by

bacterial cell wall antigens of antibody coated latex particles

The polymerase chain reaction (PCR): The high sensitivity of PCR allows detection of bacterial DNA even when concentrations are low

.Conventional assays are being replaced by newer ''real-time" system, which is faster and associated with lower contamination rates because amplification and detection occur simultaneously in a closed system

The real-time PCR is based on the measurement of a fluorescent signal generated during each amplification cycle. It produces quantitative results within 30minutes and calculates bacterial load

Broad range real-time PCR: to distinguish bacterial septicaemic dis-ease from other causes of neonatal illness such as asphyxia or compli-cations of prematurity

Red Flag Signs In A Neonate

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This article is mainly for the use of practicing pediatricians/primary care doctors and trainees, who will be reviewing babies in the neonatal period. The knowledge about the red flag signs will help you to detect or not to miss serious problems in the busy hospital/clinic practice.

Appropriate history from the mother plays a vital role while interpreting any clinical signs.

Newborn Examination:

Head to toe examination by a trained doctor or pediatrician is a must for all babies after delivery. Always assess the general well being of the baby by looking colour,tone, activity, posture , weight of the baby. All the vitals should be recorded.

Certain areas need special attention.

- Head - please look for micro/macrocephaly(confirm with HC plotting)
- Red reflex- Absence of red reflex/white reflex -indicates congenital cataract.
- O2 saturation: Always do saturation check, right hand(pre ductal) / foot (post ductal) - normal is >95. Routine pulse oxymetry is an effective screening test to detect critical CHD(clinical examination plus pulse oxymetry significantly increases sensitivity to detect CHD)
- Cleft palate- Minor clefts are usually missed unless we specifically look for and palpate.
- Spine- Skin defects, tuft of hair, dimple/pit - look for the lower end under good illumination.
- Hip: Examination to rule out developmental dysplasia of hip.
- Femoral: Feeble/absent pulsation indicates critical congenital heart disease and needs further urgent cardiac evaluation.
- Anogenital examination- Look for undescended testis, hypospadis, ambiguous genitalia, anal patency and any recto vaginal fistula.

Cardio respiratory system- Alarming symptoms and signs-

Look for symptoms like-fast breathing, recession, grunting, nasal flaring, head bobbing, apnoea, cyanosis, mottled skin, feeding difficulty, fatigue, excessive sweating, poor weight gain, pallor (please note features of heart failure may present only after first week). Also look for any signs like tachypnoea,bradycardia, prolonged capillary refill time, heart murmur and hepatomegaly.

Heart Murmur:

Normal newborn examination and absence of murmur does not guaranty that no heart disease. Many infants with murmurs do not have structural heart lesions and CHD occurs in infants without heart murmur.

Evaluation of a heart murmur is important because of potentially adverse outcomes when serious CHD remains undetected. Remember that CHD is the most common serious congenital disorder in newborns.

- Red Flags: Diastolic murmurs, Continuous murmur (especially after 48hrs by which PDA usually close), loud murmur +/- thrills, central cyanosis, tachypnea, hepatomegaly, feeble or non palpable femoral pulse.
- CCHD: Beware of Terrible T's: Tetralogy of fallot,TGA,Tricuspid Atresia, Truncus, TAPVC: All need urgent admission/ ECHO/Paediatric cardiology referral.

Respiratory distress

Any baby with respiratory distress especially grunting/apnoea needs urgent hospital evaluation and timely management.

Gastrointestinal system

Common danger signs and symptoms are refusal to feed, abdominal distension, severe/prolonged jaundice, poor weight gain, projectile/blie or blood stained vomiting, bleeding from any site.

Prolonged Jaundice

Significant jaundice persisting more than 2 weeks in term and 3 weeks in preterm babies. It can be conjugated(>15% of total) or unconjugated. Unconjugated is more common. Actively look for prolonged jaundice. Pale stools and dark urine are always alarming (Yellow alert!)

Prolonged jaundice needs clinical evaluation, appropriate investigation and management. There are treatable conditions in causes. Early identification of liver disease/extrahepatic biliary atresia improves outcome.

Vomiting

Projectile vomiting in a hungry and well baby needs to be evaluated for CHPS, especially if presentation is after 2 weeks of age. Vomiting of blood/bile always indicates a serious pathology.

Poor Weight Gain

Term babies should regain birth weight by day 10 and late Pre terms by day 14.

Average weight gain per day is: 1% of body weight

More than 10% weight loss or if weight gains less than 20 grams/ day, it needs evaluation and appropriate management. All babies' growth parameters should be plotted in an appropriate growth chart.

Common Causes to be looked into

'Not enough' : not getting enough, not taking enough, using more than baby gets, losing more than baby gets.

Feeding history is very important. If there is poor weight gain even after appropriate feeding, further evaluation is needed.

Hyper/hypothermia in a Neonate

Any neonate with recorded fever needs urgent clinical evaluation in a hospital set up. Always rule out serious infections especially if the baby is unwell. Hypothermia is an equally alarming sign.

Other associated symptoms of suspected infection are irritability, lethargy, unconsolable cry, poor/refusal to suck. System wise examination is important to find out the focus of infection. Always look for poor perfusion, respiratory distress, bulging AF, joint swelling.

Neuro-Metabolic

The commonest emergency involving CNS is neonatal seizures.

Neonatal seizure

Symptoms- They can present with a variety of symptoms like up rolling of eyes, cyclical movements of limbs, lip smacking, tonic posturing and even apnea. So in suspected cases, always take a detailed history and evaluate further for confirmation and to find the etiology and appropriate management. Don't miss to check GRBS. Video recording of episodes by parents will be useful. Please note subtle seizures are common in neonatal period.

Inborn errors of metabolism

Suspect IEM if any in baby presenting with persistant vomiting, poor weight gain, hypoglycemia,lethargy, seizures and organomegaly. They need detailed evaluation and hence referral to higher centre.

Conclusion

In our day to day life with busy clinical practice, it is always good to keep a checklist of red flag signs and symptoms while reviewing newborn babies in OPD or in hospital. This will help not to miss or early pick up of serious neonatal problems. It is very important in the present scenario of increasing medico-legal cases. Moreover to that it will help to reduce neonatal morbidity and mortality.

Pre Transport Stabilisation of a Neonate

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STABILISE BEFORE TRANSPORT

- Adequacy of Oxygenation
- Circulation
- Thermoregulation
- Acid base balance
- Metabolic control

PRE TRANSPORT CHECKLIST:

- Secure airway
- Assess adequacy of ventilation
- Assess IV access sites (if needed, reserve access sites)
- Insert NG tube to decompress stomach
- Check BP and BS
- Baby well covered, insulated and warm
- Adequate analgesia and sedation
- Collect all relevant maternal data
- Inform destination

TRANSPORT MEDICATION CHECKLIST:

- Adrenaline: 1:10,000
- Normal saline, Dextrose 10%
- Phenobarbital/ Phenytoin
- Morphine/Midazolam
- Dopamin, Prostin Infusion (in case of Cardiac newborn)
- Keep all medications ready to use(prefilled syringes)

TRANSPORT PROTOCOL

S.T.A.B.L.E. :- First introduced in US and Canada, has grown internationally to include instructor training courses in more than 45 countries.

S – Sugar, safe care

T – Temperature

A – Airway

B – Blood pressure

L – Lab work

E – Emotional support

Neonatal Transport

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Inter hospital transport should be considered if the medical resources or personal needed for a high risk neonate are not available at the hospital currently providing care. Ideally, the pregnant women should be transferred before delivery to a high risk perinatal center, when a problem is known or arises in early labor.

Indications for neonatal transfer:-

1. Extreme prematurity
2. Birth weight<1500g
3. Respiratory distress syndrome when surfactant/ ventilator is not available.
4. Severe respiratory distress with cyanosis not improve with 8cm of CPAP and ventilator is not available.
5. Meconium aspiration syndrome with PPHN, not responding to routine management.
6. Seizures not controlled with fast dose of 2 drugs.
7. Critical level of jaundice with no exportion of exchanged transfusion.
8. In born error of metabolism
9. Congenital heart disease or cardiac arrhythmia requires cardiac services
10. Surgical condition like diaphragmatic hernia, tracheoesophageal fistula, aneroid anemia.
11. Hypoglycemia not corrected with available management
12. Perfusion not improve with adequate bolos and ianotropes
13. Oliguria not responding to fluid and furosemide
14. Severe hypoxic ischemic injuries.

Steps involved in transport:

1. Communication
   a) To parent
   b) To referral center
2. Stabilization before transfer
   a) Temperature - maintenance
   b) Oxygen - prevent hypoxia
   c) Perfusion -

" IV access

" IV fluids / Bolos

" Inotropes

d) Antibiotics - first day

e) Gluon - prevent hypo/hyper glycaemia

3. Documentation

a) Antenatal

b) Immediate clinical care and management dose

c) Consent

Transport Team:-

1. 2 or 3 trainees personal in NRPand transport
2. Neonatology or pediatric trained in NRP and transport.

a) Transport equipment

Transport incubator equipped with monitors for heart rate, vascular pressures, oxygen saturation, temperature

Suction device

Infusion pumps

Gell - filled mattress

Adapters to plug into both hospital and vehicle power

Airway equipment

Anesthesia bag with manometer

Laryngoscopes with no. 0 and no. 1 blades

Magill forces

Instrument tray for chest tubes and vascular catheters

Stethoscope

Tanks of oxygen and compressed air oxygen, compressed air, light, and a source of electrical power

b) Supplies used by transport teams
c) Medications used on transport

Steps of transport :-

1. Introduction
2. Documentation before transfer
3. Consent after explains risk
4. Stabilisation if needed
5. Continues monitor during transfer
6. Documentation drugs transport
7. On arrival to NICU :-

a) Proper handover of :

- Neonate
- Documents

Special conditions and therapies during transport

1. Congenital diaphragmatic hernia

a) Intubation
b) Ventilation
c) NG tube insertion

2. Cyanotic heart disease

a) PGE1 must be available

- Ventilation if nessasary

3. Anemia

a) Blood transfusion inetiated

4. Abdominal wall defect

a) Wraping exposed contents with soaked saline

5. Tracheoesophageal fistula and esophageal atresia

a) Replogle tube
b) Continues suction

6. Neural tube defects

a) Wrapped in soaked saline

7. Severe respiratory distress with preterm

a) Surfactant administration before transfer

8. Air transport

a) Changes in barrow metric pressure/FiO2
b) Gas expantion

- Drain

o neumothorax
o Gasses distention of stomach.

Procedures in NICU

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Umbilical Vascular Access (Arterial & Venous)

Background

Umbilical vessel catheterisation is possible until the cord separates but is most successful in the first hours of life.

Access is usually under strict sterile precautions although umbilical vein catheterisation can be used for central venous access in an emergency situation.

Umbilical arterial catheterisation should be considered for any baby with increasing oxygen requirements, needing accurate blood gas monitoring, regular blood sampling or when continuous blood pressure monitoring is required.

Umbilical Artery Catheterisation (UAC)

Indications:

- Extreme Prematurity
- Increasing oxygen requirement over 40%
- Ventilated baby
- Regular blood sampling
- Invasive blood pressure monitoring
- Exchange transfusion

Contraindications to UAC

- Evidence of vascular compromise to lower limbs or buttocks
- Necrotising enterocolitis (NEC)
- Omphalitis (UVC also contraindicated)
- IUGR infants with antenatal absent or reversed end diastolic flow consider peripheral arterial line as first choice

Complications of UAC

- Sepsis
- Embolisation from air or blood clotThrombosis, which may involve:

  Femoral artery lower limb ischaemia,
  Renal artery hypertension, haematuria, renal failure,
  Mesenteric artery gut ischaemia, NEC
- Haemorrhage due to accidental disconnection

Umbilical Venous Catheterisation (UVC)

- Emergency venous access
- Central venous access for maintenance intravenous fluids, hypertonic fluids/drugs, TPN, blood products
- Exchange transfusion

Equipment:

- Sterile gown and gloves
- Umbilical sterile instruments pack
- Drapes/sterile pack and gauze
- Cord tie
- Blade / cord cutting scissors
- Silk suture with curved needle
- Umbilical arterial catheter size 2.5 F , 3.5 F
- 4F / 5F Umbilical venous catheter (Single / double lumen)
- 5 ml syringes and needle, 0.9% Saline ampoules
- 2 x 3 way taps
- Red and blue bionectors to mark UA / UV
- Fixation device/tape

Calculation for insertion lengths:

UAC = 3 x weight + 9cm + stump or diagonally umbilicus to shoulder tip length + 1cm + stump

UVC = 1.5 x weight + 5cm+ stump or {(3 x weight + 9cm + stump)/2} / 2

Emergency UVC access 5cm plus cord length

The umbilicus contains two arteries and a vein. The vein connects with the portal vein and then the vena cava. When a catheter is inserted into the umbilical vein,for emergency use, it should be in around 5cms plus cord length with easy blood aspiration.

Permanent catheter placement in the vena cava above the liver is preferred. The two small arteries direct downward on the inner aspect of the abdominal wall to connect with the left and right internal iliac arteries in the pelvis.

The UA catheter should be placed in the aorta above the diaphragm at T6-9 level or just above the bifurcation of the aorta at L 3-4 for low position placement.

Technique:

- Use sterile technique.
- Wash hands, put on sterile gown and gloves, open sterile packs,

- Prime catheter and 3-way tap with saline, leaving syringe attached throughout the procedure.
- Lift cord using sterile gauze (or ask assistant using cord clamp/forceps) clean the umbilical stump and 3-4cms of surrounding skin with 0.05% chlorhexidine solution.
- Apply sterile drapes to area, Place suture around base of cord and tie loosely to prevent excess bleeding from vessel when cut
- Holding the cord between medium forceps cut the cord cleanly using the lower edge of the forceps as a guide, leaving 1-2cm stump
- Inspect vessels and identify the arteries, smaller and thicker walled, inferior to the single vein, often standing prominent from the cut cord. Apply 2 forceps to opposite edges of the cord to stabilise and expose the vessels.
- Using a fine dilator or fine forceps gently ease the vessel open and cannulate the vessel towards the lower body (gentle upward traction of the cord may help)
- Apply gentle, steady pressure to insert the catheter. Some resistance may be felt at the umbilical ring.
- Excess force can result in a false passage.Aspirate to ensure a 'flashback' of arterial blood from the UAC, with pulsation of blood/saline present. Insert the Umbilical Venous Catheter into the vein to desired length and ensure the line samples and flushes.
- Suture the catheters separately and fix in place ensuring all connections are tight
- Ensure and record perfusion of the lower limbs once procedure completed.Infuse 1ml/hr 0.9% saline to maintain patency.
- Confirm line sites with an abdomen and chest X-Ray. Lines can be withdrawn or replaced but not advanced.
- Line positions and any adjustments must be recorded in baby's notes.
- The line length should also be marked near the stump to ensure any line displacement is quickly noted Lower limb discoloration/significantly reduced perfusion lasting over 15 minutes means line removal is indicated.

Umbilical line placement sites:

UVC:

Optimal - above diaphragm T8-10

Acceptable - Low position in IVC

Withdraw if in heart or diverted angle into liver, re Xray

UAC:

Optimal T6 - T9

Acceptable low position L3 - 4

Withdraw if at T12 - L2 as site of mesenteric and renal arteries

Line Removal:

UAC:

To remove an arterial umbilical catheter, stop the heparin infusion but continue monitoring

pulsation trace. Under sterile precautions withdraw the catheter to 5cm length then withdraw by 1cm per minute until the arterial trace has completely flattened. Remove catheter and put pressure on the artery to stop any bleeding.

UVC:

To remove a venous umbilical catheter, stop any infusion,

withdraw the catheter until it is just outside of the abdominal wall. Wait for clotting, then remove the catheter entirely. Be careful to occlude the vein because air embolism may result if the vessel remains open.

Pneumothorax

BACKGROUND

A Pneumothorax may be an emergency when the air collection is under pressure (a tension pneumothorax). When it causes an acute clinical deterioration it may be necessary to drain the pneumothorax by needle aspiration and/or chest drain insertion

PURPOSE

To drain air from the pleural cavity allowing the lung to re-inflate thus improving baby's condition/ventilation.

MAKING THE DIAGNOSIS

Suspect a pneumothorax if

- increase in respiratory distress and/or diminished chest movements
- sudden deterioration with desaturation
- circulation may become compromised blood gas shows hypoxia, respiratory and/or metabolic acidosis.

Clinical signs

- May be minimal unequal or decreased air entry
- Asymmetrical chest movements
- tachycardia
- fall in blood pressure
- transillumination with a cold light. Useful but can be unreliable (esp in extreme preterms and babies with chest wall edema)
- CXR will confirm the diagnosis but in an emergency is usually too time consuming.

Needle Aspiration of Chest

Needle aspiration is an emergency procedure only. Care must be taken to avoid laceration of the lung or puncturing blood vessels.

Equipments:

- 21 gauge (green) or 23 gauge (blue) butterfly needle
- 3 way tap
- 10 ml syringe
- Alcohol skin wipe
- 1 pair sterile gloves

Procedure

- Infant supine, prepare area with alcohol wipes
- Insert needle into the pleural space (directly over the top of the rib in the 2nd or 3rd intercostal space in the mid-clavicular line) until air is aspirated into the syringe, then expel air through the 3-way stopcock.

Ongoing Care

Following needle aspiration, insertion of an intercostal catheter is usually required for on-going management.

Catheter - Trocar Chest Drain Insertion

EQUIPMENT:

Sterile chest drain pack (Size 8/10) with trochar
Sterile gloves, gown and drapes
2% Xylocaine solution for LA
Size 11 Pointed tip blade for skin incision
3/0 Centisilk for suturing
Syringes
Adhesive tapes

PROCEDURE

- Inform parents where possible
- Sterile gown and gloves
- Aim to maintain the infant's temperature. Place the infant with the affected side uppermost and the arm extended above the head. Ensure limbs are adequately restrained.
- Monitor infant's heart rate and oxygen saturation level
- The intercostal catheter ("ICC", "chest drain") is usually inserted in the 4th or 5th intercostal space in the mid-axillary line. This corresponds to a point 1-2cm lateral to and 0.5-1cm below the nipple.
- The incision must be well clear of the nipple.
- Mark location with pen.
- Prepare the field with 0.05% chlorhexidine (pink solution)
- Select intercostal catheter size

Infants

> 1500g -- 10 or 12 Fr
< 1500g -- 8 or 10 Fr
<1000g -- 8 Fr

- Place sterile drape in position
- Infiltrate the insertion site with 1% Lignocaine 0.5 - 1mL (max). If baby is ventilated and on a morphine infusion can also give a bolus dose of 100 micrograms per kg, which can be repeated if needed.
- Using small scalpel blade make a 1cm incision through the skin and subcutaneous tissue
- Using straight mosquito forceps to bluntly dissect away the subcutaneous tissue and intercostal muscles, the parietal pleura is reached. Aim to dissect a passage just above a rib border in order to avoid the neurovascular bundles running below each rib.
- Open the parietal pleura by blunt dissection. At this point the hiss of air escaping the pleural space may be heard
- Remove the trocar from the ICC and grasp the distal end with the curved artery forceps. Direct the tip anteriorly as well as superomedially so that the tip lies beneath the anterior chest wall. Advance the ICC into the pleural space 2 - 4 cm, depending on the baby's size.
- Connect the ICC via connector to an underwater seal drainage system (Sentinel Seal), and note whether the fluid is swinging and/or bubbling. Condensation within the catheter may be seen when within the pleural space.
- Place a single stitch through the wound so that the skin is drawn snugly around the ICC. Purse string stitches are not used as they leave an unsightly scar. Wrap the ends of the suture around the ICC several times and tie securely.
- Secure the ICC to the chest wall with Tegaderm. Position it to maintain the anterior position of the ICC. Secure positioning is important to minimize trauma to intrathoracic structures due to movement of the extrathoracic portion of the ICC.

Ongoing Care

- The need for ongoing analgesia is based on an assessment of physiological and behavioural responses associated with pain.

Instructions for removal

- The decision to clamp off and/or remove a chest drain should be considered once no reaccumilation is ensured / baby is off positive pressure ventilation.

Intraosseous access

Intraosseous (IO) access is an effective route for fluid resuscitation, drug delivery and laboratory evaluation that may be attained in all age groups and has an acceptable safety profile.

Indications

- IO access is the recommended technique for circulatory access in cardiac arrest.
- In decompensated shock IO access should be established if vascular access is not rapidly achieved (if other attempts at venous access fail, or if they will take longer than ninety seconds to carry out.)
- The exception is the newborn, where umbilical vein access continues to be the preferred route.

Contraindications

- Proximal ipsilateral fracture
- Ipsilateral vascular injury
- Osteogenesis imperfecta

Complications

- Failure to enter the bone marrow, with extravasation or subperiosteal infusion
- Through and through penetration of the bone
- Osteomyelitis (rare in short term use)
- Physeal plate injury
- Local infection, skin necrosis, pain, compartment syndrome, fat and bone microemboli have all been reported but are rare

Equipment

- Alcohol swabs
- 18G needle with trochar (at least 1.5 cm in length)
- 5 ml syringe
- 20 ml syringe
- Infusion fluid

Analgesia, Anaesthesia, Sedation

Local anaesthesia may be required if the patient is conscious.

Procedure

- Identify the appropriate site
- Proximal tibia: Anteromedial surface, 2-3 cm below the tibial tuberosity
- Distal tibia: Proximal to the medial malleolus
- Distal femur: Midline, 2-3 cm above the external condyle
- Prepare the skin
- Insert the needle through the skin, and then with a screwing motion perpendicularly / slightly away from the physeal plate into the bone. There is a give as the marrow cavity is entered
- Remove the trocar and confirm position by aspirating bone marrow through a 5 ml syringe.
- Marrow cannot always be aspirated but it should flush easily.
- Secure the needle and start the infusion (this needs to be manually administered as boluses with the 20 ml syringe.

Laboratory tests

Most laboratory tests cannot be performed on aspirated bone marrow as the particulate matter may block and damage laboratory equipment

For urgent transfusion support in the absence of a pretransfusion blood sample (not bone marrow) - universal donor products (Group O blood cells, Group AB plasma) will be issued

Aspirated bone marrow is suitable for blood culture bottles, bedside glucometers and point of care devices

Post-procedure care

Intraosseous infusion should be limited to emergency resuscitation of the child and discontinued as other venous access has been obtained.

PERICARDIOCENTESIS

- Pneumopericardium is the collection of air in the pericardial space
- Pericardial effusion is the accumulation of excess fluid in the pericardial space
- Pericardiocentesis is a procedure to remove air or excess fluid from the pericardial space, usually through a needle, small cannula, or drainage catheter.

Indications

1. Cardiac tamponade due to pneumopericardium
2. Cardiac tamponade due to pericardial fluid
3. Aspiration of pericardial fluid for diagnostic studies

Contraindications

- There are no absolute contraindications to performing pericardiocentesis in the setting of cardiac tamponade.
- Relative contraindication for diagnostic pericardiocentesis
- Coagulopathy
- Active infection. (However, infection may also be an indication for diagnostic pericardiocentesis in some clinical situations.)

Equipment

- Sterile field with aperture drape or multiple drapes to be arranged around access site
- Sterile swabs or gauze pads
- Sterile gloves
- Local anesthetic, as needed
- 16- to 20-gauge intravenous cannula over 1- to 2-in needle
- Indwelling drainage catheter (optional)
- Three-way stopcock

- Short intravenous extension tubing (optional)
- 10- to 20-mL syringes
- Preassembled closed drainage system as for Emergency Evacuation of Air Leaks, Thoracostomy Tubes (optional)
- Connecting tubing and underwater seal for indwelling drain (optional)
- Transillumination device (optional, for pneumopericardium)
- Echocardiogram/sonography imaging device (optional in urgent situations)
- Specimen containers for laboratory studies, if procedure is diagnostic

Precautions

- Draining a large volume from the pericardial space can alter cardiac preloading conditions significantly, and some infants may require a supplemental intravascular fluid bolus after the pericardium is drained.

Techniques

- If ultrasound/echocardiographic imaging is available, and if time permits, then imaging can be performed to determine an optimal entry site and angle. In addition, the approximate distance required to reach the pericardial space can be estimated.
- Similarly, evaluation with transillumination can be performed in cases of pneumopericardium, if time permits.
- Cleanse skin over xiphoid, precordium, and epigastric area with antiseptic. Allow to dry.
- Arrange sterile drapes, leaving the subxiphoid area exposed.
- Local anesthesia should be administered for the conscious patient. A typical example is 0.25 to 1.0 mL of subcutaneous 1% lidocaine instilled within 1 to 2 cm of the xiphoid process.
- Assemble the needle/cannula, three-way stopcock, and syringe so that the stopcock is open to both the needle and the syringe, but closed to the remaining port.
- The usual entry point in an infant is 0.5 to 1 cm below the tip of the xiphoid process, in the midline or slightly (0.5 cm) to the left of the midline. The needle should be elevated 30 to 40 degrees at the skin, and the tip should be directed toward the left shoulder. A different approach may be used in certain cases, for example, if an echocardiogram suggests that most of the fluid is right-sided or apical.
- While advancing the needle, apply gentle negative pressure with the syringe. Continue advancing until air or fluid is obtained. If the syringe fills, use the third port of the stopcock to empty the syringe, or to attach a second syringe, and then aspirate more, repeating as needed. If diagnostic studies are desired, the fluid should be transferred to appropriate specimen containers.
- If bloody fluid is aspirated, there could be a serosanguineous or hemorrhagic effusion, or the needle might have entered the heart (usually the right ventricle).
- A rhythmic tug, corresponding to the heart rate, may be felt as the needle enters the pericardium. Although this tugging sensation can reflect entering the myocardium, it can also be felt while the tip of the needle is positioned correctly within the pericardial space, and it does not necessarily mean that the needle has entered the heart.
- If ultrasound imaging is available, needle position can be determined either by visualizing the tip of the needle within the pericardial space or by demonstrating that the amount of pericardial fluid is diminishing as fluid is aspirated.
- Once the needle is in the pericardial space, as much pericardial fluid or air should be evacuated as possible. To accomplish this, fix the needle in position and advance the cannula over the needle into the pericardial space. Remove the needle, and connect the cannula to a closed system for aspiration, such as a three-way stopcock and a syringe. Aspirate as much fluid/air as possible.
- Note that small single-lumen catheters may easily become blocked.
- A decision will need to be made whether to leave the cannula in place for any length of time or to remove it once the pericardium has been drained. This decision will vary in individual cases, but factors to consider include the likelihood of reaccumulation and need for repeat drainage versus the risk of infection or entry of free air with an indwelling cannula.
- In certain cases, the operator may elect to evacuate the pericardial space directly through the needle, rather than placing a cannula.
- Draining a large volume from the pericardial space can alter cardiac preloading conditions significantly, and some infants may benefit from intravascular fluid boluses after the pericardium is drained.
- Pericardiocentesis is often an urgent or emergency procedure. The technique for pericardiocentesis described above applies when there is time for each step. In an infant with significant hemodynamic compromise, the operator may be forced to omit certain steps in the interest of time.

Complications:

Pneumopericardium

Pneumomediastinum

Pneumothorax

Cardiac perforation

Arrhythmia

Hypotension (if a large effusion is drained)

Charts and tables in NICU

Dr. Shabeer M P[1], Dr. Nihaz Naha K[2]

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GUIDELINES FORFLUID REQUIREMENTS

Day	1	2	3	4	5+
Ml/kg/day	60	80	100	120	140

Calculate on the greater of birth or actual weight, provided the infant is not oedematous. Preterm babies, start at 80 ml/kg/day.

This requirement is inclusive of insensible water loss, urine output and stool output.

- Fluid adjusted depending on weight loss.
- Initial fluid D 10% on day 1 and day 2 followed by cocktail with Na and K+ depending on electrolyte report.
- Initially baby lose weight about 1-3% per day to a maximum of 10-15% of birth weight.
- Fluid loss other than mentioned above is replaced usually with 0.45 saline with or without K+ every 4 - 8 hours.

The choice of fluid depends on the site of fluid loss.

Site	Fluid replacement
Gastric	NS
Duodenal/Jejunal	½ NS with K+
Ileal	½ NS with K+
Colon	NS
Renal	Fluid without K

FLUID CALCULATION

Formula to calculate IV fluids/ Dextrose

- How to calculate IV dextrose when the required concentration eg 15%

X = Z (C - B / A - B)

- X - Volume of solution A (10% Dextrose) needed
- Y - Volume of solution B (50% OR 25% Dextrose) needed

- Z - Volume of desired solution &
- C - the desired %
- Eg X = 100 (15 - 50 / 10 - 50) = 100 (- 35/ - 40) = 100 (35/40) = 87.5 ml of 10% Dextrose
- 100 - 87.5 = 12.5 ml of 50% Dextrose
- 8.75 + 6.25 g = 15 %

Y = Z - X

SOME QUICK RESPONSE EXAMPLES

12% D	43ML 10% D + 7ML 25% D
15% D	33ML 10% D + 17ML 25% D
18% D	24ML 10% D + 26ML 25% D
20% D	18ML 10% D + 32ML 25% D

How to check GIR (Glucose Infusion Rate)

Eg:- 10 x 100 / 144 = 6.2 mg / kg / min

Eg:- 0.166 x 10 x 10 / 2 kg

How to calculate Dextrose volume when two solutions are involved (N/5 with desired percentage of Dextrose

X = C (TV) - (Dex V) B / (A - B)

- X - Volume of solution A (10% dextrose) needed
- A = 10% Dextrose
- B = 50% Dextrose
- TV = Total volume of desire solution &
- C is the desired %
- Dex Vol - TV - Vol of other fluids like saline, amino acid, KCL etc
- Vol of 50% dextrose (B) = dextrose vol - X
- Eg : Total volume (TV) = 100 ml, Other fluids = 30 ml, Solution needed is 10 % Dex
- X = 10 (100) - (70) 50 / 10 - 50
  X = 1000 - 3500 / - 40
  X = 250/4 = 62.5 ml of 10% Dextrose, so 50% = 7.5 ml(70-62.5)
  (6.25+3.75 = 10 g / 100 ml = 10 % solution)

Table 17.2 Recommendations for intravenous mineral, trace elements and vitamins in very low birthweight infants (amount per kilogram per day)

Sodium	3–5 mmol
Chloride	3–7 mmol
Potassium	2–3 mmol
Calcium	1.5–2.0 mmol
Phosphorus	1.5–1.9 mmol
Magnesium	0.2–0.3 mmol
Zinc	6.1 µmol
Copper	0.3 µmol
Selenium	19–57 nmol
Manganese	18.2 nmol
Iodine	7.9 nmol
Chromium	1–5.8 nmol
Molybdenum	2.6 nmol

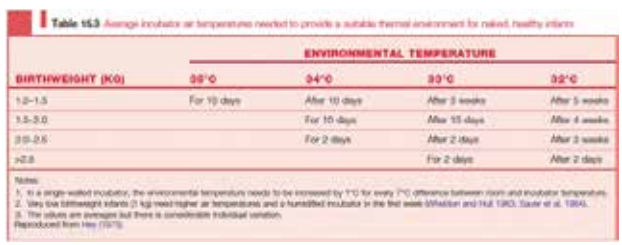
Table 17.3 Vitamins

RECOMMENDATIONS		COMPOSITION of VITLIPID (per ml)
Vitamin A	700–1500 IU	230 units
Vitamin D	40–160 IU	40 units
Vitamin E	2.8–3.5 IU	0.7 units
Vitamin K	10 µg	20 µg

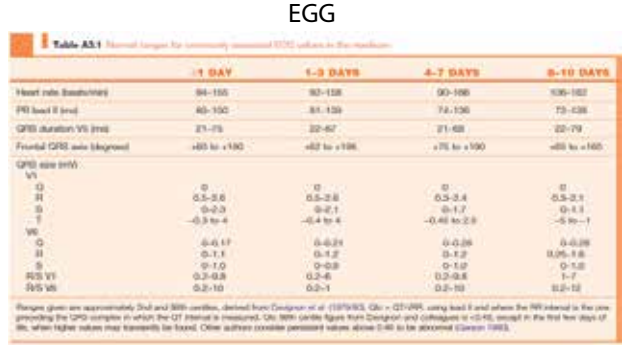
Temperature

Table 15.4 Suggested abdominal skin temperature settings for infants nursed under radiant warmers or in servo mode incubators

BIRTHWEIGHT (kg)	ABDOMINAL SKIN TEMPERATURE (°C)
<1.0	36.9
1.0–1.5	36.7
1.5–2.0	36.5
2.0–2.5	36.3
>2.5	36.0



EGG



Blood Pressure

Table A5.1 Normal ranges for commonly measured ECG values in the newborn

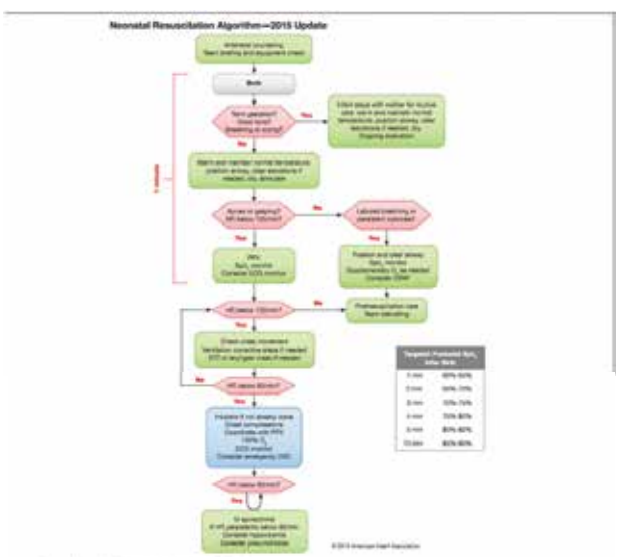
	1 DAY	1–3 DAYS	4–7 DAYS	8–10 DAYS
Heart rate (beats/min)	94–155	92–158	90–166	106–162
PR (and II) (ms)	40–150	81–139	74–136	73–138
QRS duration V6 (ms)	21–75	22–67	21–68	22–79
Frontal QRS axis (degrees)	+85 to +190	+62 to +196	+75 to +190	+85 to +165
QRS size (mV)				
V1				
Q	0	0	0	0
R	0.5–3.6	0.5–2.6	0.5–3.4	0.5–2.1
S	0–2.3	0–2.1	0–1.7	0–1.1
T	−0.3 to +4	−0.4 to +4	−0.46 to 2.9	−5 to −1
V6				
Q	0–0.17	0–0.21	0–0.26	0–0.28
R	0–1.1	0–1.2	0–1.2	0.05–1.6
S	0–1.0	0–0.9	0–1.0	0–1.0
R/S V1	0.2–0.8	0.2–6	0.2–0.8	1–7
R/S V6	0.2–10	0.2–1	0.2–10	0.2–12

Ranges given are approximately 2nd and 98th centiles, derived from Davignon et al. (1979/83). Qtc = QT/√RR, using lead II and where the RR interval is the one preceding the QRS complex in which the QT interval is measured. Qtc 98th centile figure from Davignon and colleagues is <0.46, except in the first few days of life, when higher values may transiently be found. Other authors consider persistent values above 0.46 to be abnormal (Gerson 1983).

Table A4.2 Estimated BP values after a variable of age in weeks from 26 to 44 weeks postmenstrual age (from Dionne et al. 2011, with permission)

POSTCONCEPTIONAL AGE	50TH PERCENTILE	95TH PERCENTILE	99TH PERCENTILE
44 weeks			
SBP	88	106	110
DBP	55	68	73
MAP	63	80	85
42 weeks			
SBP	85	98	102
DBP	53	65	70
MAP	62	76	81
40 weeks			
SBP	80	95	100
DBP	50	63	70
MAP	60	75	80
38 weeks			
SBP	77	92	97
DBP	50	65	70
MAP	60	74	78
36 weeks			
SBP	72	87	92
DBP	50	65	70
MAP	57	72	74
34 weeks			
SBP	70	85	90
DBP	50	65	60
MAP	60	65	70
32 weeks			
SBP	68	83	88
DBP	42	56	60
MAP	49	62	68
30 weeks			
SBP	65	80	85
DBP	43	55	60
MAP	45	66	68
28 weeks			
SBP	60	75	80
DBP	39	50	54
MAP	48	58	63
26 weeks			
SBP	55	72	77
DBP	30	50	56
MAP	38	57	63

NRP 2015



BLOOD INDICES

Table A2.1 Reference values for coagulation tests in healthy term infants and in the adult (means and standard deviations)

TEST	DAY 1	DAY 5	DAY 30	ADULT
PT (s)	13 ± 1.43	12.4 ± 1.46	11.8 ± 1.25	12.4 ± 0.76
APPT (s)	42.9 ± 5.8	42.9 ± 8.62	40.4 ± 7.42	33.5 ± 3.44

Table A1.1 Red cell parameters in the fetus

AGE (weeks)	Hb (g/dl)	PCV	RBC ($\times 10^{12}$/l)	MCV (fl)	MCH (pg)	MCHC (g/dl)	NUCLEATED RBC (% OF WBC)	RETICULOCYTES (%)
12	8.0–10.0	0.33	1.5	160	60	34	5.0–8.0	40
16	10.0	0.38	2.0	140	45	33	3.0–4.0	10–25
20	11.0	0.37	2.5	135	44	33	1.0	10–20
24	14.0	0.40	3.5	123	38	31	1.0	5–15
28	14.5	0.45	4.0	120	40	31	0.5	5–20
34	15.0	0.47	4.4	118	38	32	0.2	3–10
Term 40 cord	16.8	0.53	5.35	107	34	31.7	0.01	3–7

Hb, haemoglobin; MCH, mean corpuscular haemoglobin; MCHC, mean corpuscular haemoglobin concentration; MCV, mean cell volume; PCV, packed cell volume; RBC, red blood cells; WBC, white blood cells.
Adapted from Oski, F.A., Naiman, J.L., 1982. Hematologic Problems in the Newborn, third ed. WB Saunders, New York.

Table A1.2 Normal leukocyte counts ($\times 10^9$/l) in the first month of life

AGE	TOTAL LEUKOCYTES, MEAN (RANGE)	NEUTROPHILS, MEAN (RANGE)	%	LYMPHOCYTES, MEAN (RANGE)	%	MONOCYTES, MEAN	%	EOSINOPHILS, (MEAN)	%
Birth	18.1 (9.0–30.0)	11.1 (6.0–26.0)	61	5.5 (2.0–11.0)	31	1.1	6	0.4	2
12 h	22.8 (13.0–38.0)	15.5 (8.0–28.0)	68	5.5 (2.0–11.0)	24	1.2	5	0.5	2
24 h	16.8 (9.4–34.0)	11.5 (5.0–21.5)	61	5.8 (2.0–11.5)	31	1.1	6	0.3	2
1 week	12.2 (5.0–21.0)	5.9 (1.5–10.0)	45	5.0 (2.0–17.0)	41	1.1	9	0.5	4
2 weeks	11.4 (5.0–20.0)	4.5 (1.0–8.5)	40	5.5 (2.0–17.0)	48	1.0	9	0.4	3
1 month	10.8 (5.0–19.5)	3.8 (1.0–8.0)	35	6.0 (2.5–16.5)	56	0.7	7	0.3	3

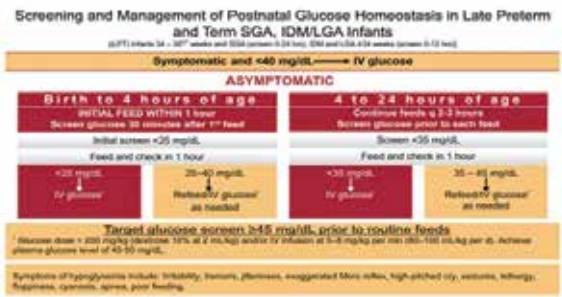
Neutrophils include band forms and a small number of metamyelocytes and myelocytes in the first few days of life.
With permission from Sellmer, P.R., 1977. Reference ranges for leukocyte counts in children. In: Rudolph, A.M. (Ed.), Pediatrics, sixteenth ed. Appleton-Century-Crofts, New York, p. 1118.

CSF REFERENCE RANGE

TYPE OF INFANT	WHITE CELL COUNT (COUNT/mm^3)	PROTEIN (g/l)	GLUCOSE (mmol/l)
Preterm <28 days	9 (0–30)	1 (0.5–2.5)*	3 (1.5–5.5)
Term <28 days	6 (0–21)	0.6 (0.3–2.0)*	3 (1.5–5.5)

All values are given as mean and range.
Table combined from a review of the literature (see reference list).
*Protein values are higher in the first week of life and depend on the red cell count. A white cell count of more than 21/mm^3 with a protein value of more than 1 g/l with less than 1000 red cells is suspicious of meningitis.

HYPOGLYCEMIA



ET Tube size chart

Tube Size (internal diameter)	Weight (g)	Gestational Age
2.5	<1000	<28
3.0	1000-2000	28-34
3.5	2000-3000	35-40
3.5-4.0	>3000	>38

Depth of insertion of ET tube

1.	wt + 6 as measured from the upperlip of neonate.
2.	distance from nasal septum to tragus(NTL) +1 as measured from the upperlip of neonate.

Table 13.1 Oral tracheal tube lengths by gestation

GESTATION (WEEKS)	TRACHEAL TUBE AT LIPS (CM)
23–24	5.5
25–26	6.0
27–29	6.5
30–32	7.0
33–34	7.5
35–37	8.0
38–40	8.5
41–43	9.0

(Reproduced from Kempley et al. (2008)).

Normal Blood Gas Values

Table A7.1 Normal blood gas values

	PaO_2				$PaCO_2$				H^+			
	kPa	mmHg	kPa	mmHg	kPa	mmHg	kPa	mmHg	nmol/l	pH	nmol/l	pH
15 min	11.6	87			3.7	28			48	7.32		
30 min	11.4	86			4.3	32			43	7.37		
60 min	10.8	81			4.1	31			40	7.40		
1–6 h	8.0–10.6	60–80	8.0–9.3	60–70	4.7–6.0	35–45	4.7–6.0	35–45	46–49	7.31–7.34	42–48	7.32–7.38
6–24 h	9.3–10	70–75	8.0–9.3	60–70	4.4–4.8	33–36	3.6–5.3	27–40	37–43	7.37–7.43	38–45	7.36–7.45
48 h–1 week	9.3–11.2	70–85	10.0–10.6	75–80	4.4–4.8	33–36	4.3–4.5	32–35	42–44	7.36–7.38	40–48	7.32–7.40
2 weeks					4.8–5.2	36–39	5.1	38	43	7.37	48	7.32
3 weeks					5.3	40	5.1	38	42	7.38	49	7.31
1 month					5.3	39	4.9	37	41	7.39	49	7.31

The values at 15, 30 and 60 minutes are from our own unpublished observations on full term infants. Data from 1 hour to 1 week are drawn from the literature on arterial samples. Data beyond 1 week are on capillary samples. Values in the table which are in italics are those for premature infants; those not in italics are for full-term infants.
Reproduced from Rennie, J.M., Roberton, N.R.C., 2002. Manual of Neonatal Intensive Care, fourth ed. Edward Arnold, London.

Ventilator adjustments based on blood gas

Table 27.12 Adjustments to ventilator settings on the basis of blood gas changes

Low PaO_2	High $PaCO_2$	Increase peak pressure, which will also increase mean airway pressure: in spontaneously breathing babies ↑ rates may also work
Low PaO_2	Normal $PaCO_2$	↑ FiO_2; ↑ MAP but maintain PIP (i.e. ↑ PEEP or ↑ T_I)
Low PaO_2	Low $PaCO_2$	Consider alternative diagnosis, e.g. PPHN, sepsis, overventilation. ↑ FiO_2; ↑ MAP; use vasodilators
PaO_2 normal	High $PaCO_2$	↓ PEEP, ↑ rate; keep MAP constant
PaO_2 normal	Low $PaCO_2$	↓ rate; maintain MAP
PaO_2 high	$PaCO_2$ high	Rare: check for mechanical problems, e.g. blocked tube. ↓ PEEP, ↓ T_I, ↑ rate ↓ FiO_2
PaO_2 high	$PaCO_2$ normal	↓ MAP (usually ↓ PIP): ↓ FiO_2
PaO_2 high	$PaCO_2$ low	↓ pressure, ↓ rate, ↓ FiO_2 (see text)
PaO_2 normal	$PaCO_2$ normal	Sit tight! Unless plan to wean

MAP, mean arterial pressure; PIP, peak inspiratory pressure; PEEP, positive end-expiratory pressure; PPHN, persistent pulmonary hypertension of the newborn.

Derived ventilation indices

Table 27.17 Derived predictive indices in congenital diaphragmatic hernia

Oxygenation index (OI)

$$OI = \frac{\text{mean airway pressure } (cmH_2O) \times FiO_2 (\%)}{PaO_2 (KPa)}$$

(NB: if PaO is more familiar in mmHg then divide result by 7.5)

Ventilation index (VI)

VI = respiratory rate (breaths/min) × mean airway pressure (cmH_2O)

Wilford Hall/Santa Rosa prediction formula

$WHSR_{PF}$ = highest PO_2 – highest PCO_2

(N.B. measured on first day)

Transfusion guidelines in newborn

Table 30.13 Guidelines for red cell transfusion for preterm neonates

ASSISTED VENTILATION			CPAP		BREATHING SPONTANEOUSLY	
<28 days		≥28 days	<28 days	≥28 days	FiO_2 > 0.21	Well in air
[illegible]	[illegible]					
Hb < 12 g/dl or PCV < 0.40	Hb < 11 g/dl or PCV < 0.35	Hb < 10 g/dl or PCV < 0.30	Hb < 10 g/dl or PCV < 0.30	Hb < 8 g/dl or PCV < 0.25	Hb < 8 g/dl or PCV < 0.25	Hb < 7 g/dl or PCV < 0.20

RBC transfusion may be considered at higher thresholds than the above for neonates with:
- hypovolaemia (unresponsive to crystalloid infusion)
- septic shock
- necrotising enterocolitis
- undergoing/recovering from major surgery.

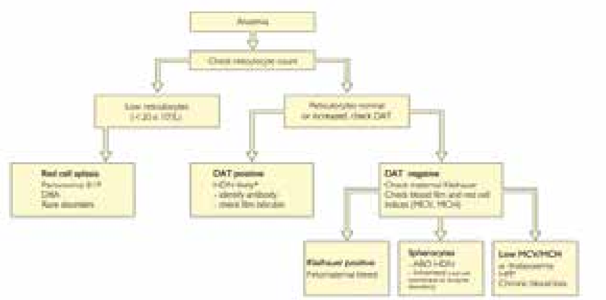
CPAP, continuous positive airway pressure; Hb, haemoglobin; PCV, packed cell volume.

Table 30.14 Guidelines for platelet transfusion for neonates

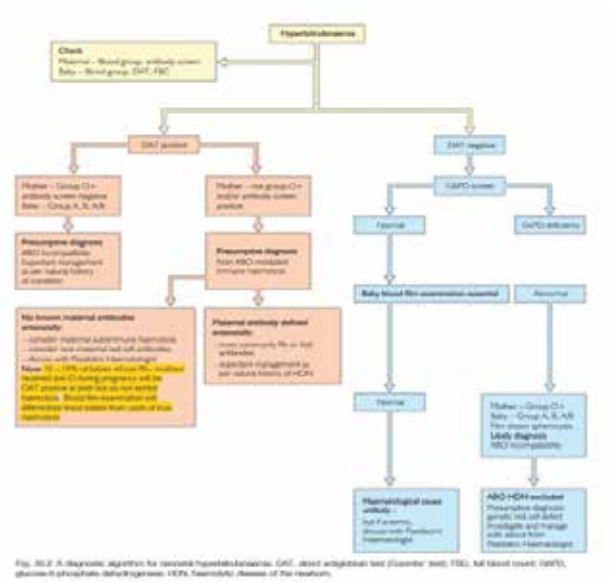
PLATELET COUNT (×10⁹/l)	NON-BLEEDING NEONATE	BLEEDING NEONATE	NAITP (PROVEN OR SUSPECTED)
<30	Consider transfusion in all patients	Transfuse	Transfuse (with HPA-compatible platelets)
30–49	Do not transfuse if clinically stable Consider transfusion if: • <1000 g and <1 week of age • clinically unstable (e.g. fluctuating BP) • previous major bleeding • [illegible] • current minor bleeding [illegible] • concurrent coagulopathy • requires surgery or exchange transfusion • platelet count falling and likely to fall below 30	Transfuse	Transfuse (with HPA-compatible platelets)
50–99	Do not transfuse	Transfuse	Transfuse (with HPA-compatible platelets if major bleeding present)
>99	Do not transfuse	Do not transfuse	Do not transfuse

NAITP, neonatal alloimmune thrombocytopenia; HPA, human platelet antigen; BP, blood pressure; [illegible]

Approach to Anemia in newborn



Approach to Hyperbilirubinemia



Causes of abnormal Coagulation tests

Table 30.10 Causes of abnormal coagulation tests in the newborn

Prolonged APTT alone	Inherited deficiency of factor VIII, factor IX, factor XI, factor XII Heparin (therapeutic or heparin contamination*)
Prolonged PT alone	Inherited deficiency of factor VII Vitamin K deficiency (haemorrhagic disease of the newborn) Warfarin
Prolonged TT alone	Low fibrinogen Artefact: contamination with heparin from line or sample bottle*
Prolonged APTT + PT	Inherited deficiency of factor II, factor V, factor X Liver disease
Prolonged APTT, PT and TT	Inherited deficiency of fibrinogen Disseminated intravascular coagulation Severe liver disease
Normal APTT, PT	Factor XIII deficiency and TT Platelet defect: thrombocytopenia or rare platelet function abnormality (e.g. Bernard Soulier syndrome)

APTT, activated partial thromboplastin time; PT, prothrombin time; TT, thrombin time.
*The effect of heparin contamination can be distinguished by checking the reptilase time on a coagulation sample; where a prolonged TT and/or a prolonged APTT is due to heparin contamination, the reptilase time will be normal, but where a prolonged TT or APTT is due to a true coagulation defect, the reptilase time will be abnormal.

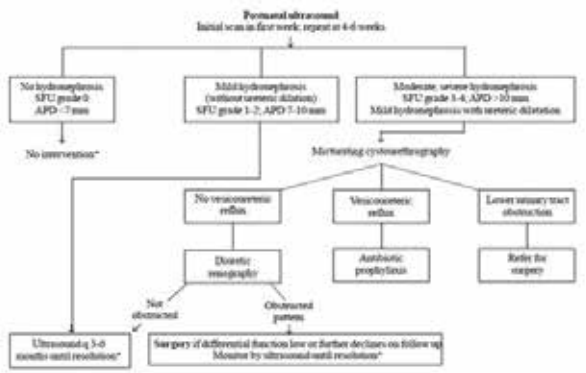
Drugs and chemicals associated with hemolysis in G6PD deficient patients

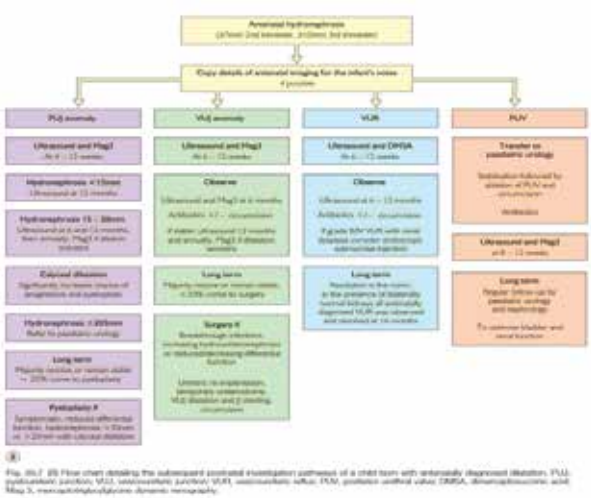
Table 30.6 Drugs and chemicals associated with haemolysis in patients who are glucose-6-phosphate dehydrogenase (G6PD)-deficient

Antimalarials
Primaquine Pamaquine (Quinine)* (Chloroquine)*
Antibiotics
Nitrofurantoin Sulphones, e.g. dapsone Sulphonamides,† e.g. sulphamethoxazole (Septrin) Quinolones, e.g. nalidixic acid, ciprofloxacin (Chloramphenicol)‡
Analgesics
Aspirin (in high doses) Phenacetin
Chemicals
Naphthalene (mothballs) Divicine (fava beans – also known as broad beans) Methylene blue

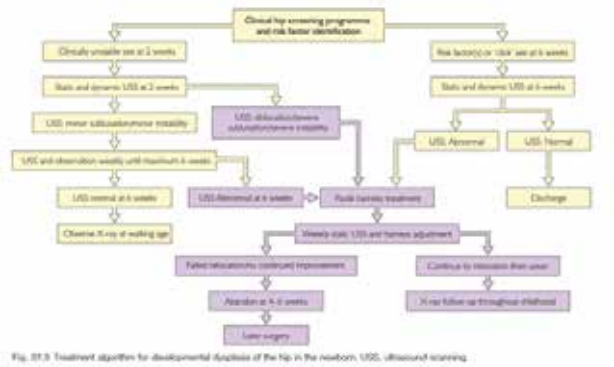
*Acceptable in acute malaria.
†Some sulphonamides do not cause haemolysis in most G6PD-deficient patients, e.g. sulfadiazine.
‡To be avoided in some types of G6PD deficiency (can be taken by patients with the common, African A-form of G6PD deficiency).

Approach to antenatal hydronephrosis

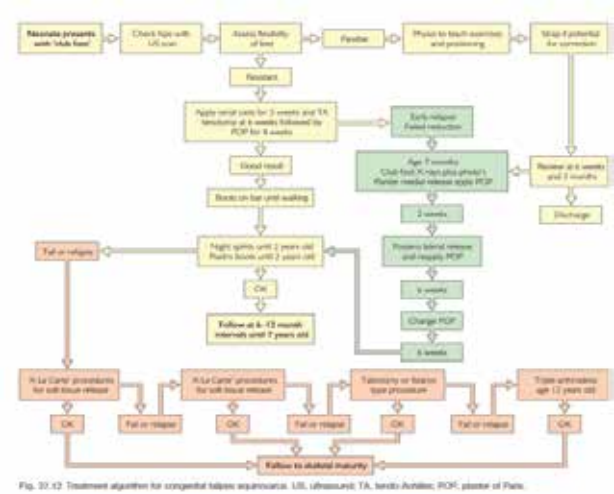




Treatment algorithm for DDH in newborn



Treatment algorithm for CTEV in newborn



Thompson score

	SCORE			
Sign	0	1	2	3
Tone	Normal	Hypertonic	Hypotonic	Flaccid
conscious state	Normal	Hyperalert, irritable	Lethargic	Comatose
Fits	Normal	Infrequent, <3/day	Frequent, >2/day	
Posture	Normal	Fisting, cycling	Strong distal flexion	Decerebrate
Moro	Normal	Partial	Absent	
Grasp	Normal	Poor	Absent	
Suck	Normal	Poor	Absent/bites	
Respiration	Normal	Hyperventilation	Brief apnoea	Apnoeic
Fontanelle	Normal	Full, not tense	Tense	

Modified Levene & Starte chart for ventricular index

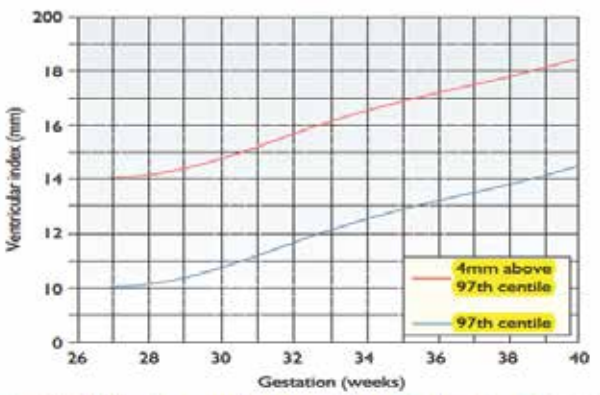


Fig. 40.52 Chart for normal ranges of the ventricular index. *(Modified from Levene and Starte 1981.)*

Emergency management of Hyperkalemia

Table 35.4 Emergency management of hyperkalaemia

Intravenous salbutamol (4 µg/kg over 5 minutes) or nebulised salbutamol (2.5–5 mg); repeated as necessary

Intravenous glucose and insulin infusion (12 units soluble insulin in 100 ml 25% glucose; 5 ml/kg given over 30 minutes); has an additive effect with salbutamol; monitor blood glucose closely during and after treatment

Intravenous sodium bicarbonate 1 mmol/kg (2 ml/kg 4.2%); effective even if infant is not acidotic

10% calcium gluconate (0.1 ml/kg) by intravenous injection over 10 minutes; repeated as necessary to maintain normocalcaemia (caution: do not administer in same line as bicarbonate; risk of precipitation)

Urinary indices in Prerenal failure and ATN

Table 35.3 Urinary biochemical indices in prerenal failure and acute tubular necrosis in term babies

TEST	PRERENAL FAILURE	ACUTE TUBULAR NECROSIS
Urine urea	High	Low
Urine creatinine	High	Low
Urine osmolality	High (>500 mOsmol/kg)	Low (>300 mOsmol/kg)
Specific gravity	High (>1.025)	Low (>1010)
Urine sodium	Low (<10 mmol/l)	High (>20 mmol/l)
U:P urea	High (>10)	Low
U:P creatinine	High (>40)	Low
U:P osmolality	High (>2)	Low (>1)
FE_{Na}	Low (<1%)	High (>3%)
RFI	Low (<1)	High (>4)

FE_{Na}, fractional sodium excretion; RFI, renal failure index. See text for calculation and interpretation in extremely preterm babies.

NUTRITION

Table 16.2 Recommended intakes of individual nutrients for formula-fed stable/growing preterm infants

	ESPGHAN		TSANG ELBW		VLBW	
	per kg/day	per 100 kcal	per kg/day	per 100 kcal	per kg/day	per 100 kcal
Energy						
kcal	110–135		130–150		110–130	
kJ						
Protein (g)	4.0–4.5 (<1 kg) 3.5–4.0 (1–1.8 kg)	3.6–4.1 (<1 kg) 3.2–3.6	3.8–4.4	2.5–3.4	3.4–4.2	
Fat (g)	4.8–6.6	4.4–6.0 (<40% MCT)	6.2–8.4	4.1–6.5	5.3–7.2	
Linoleic acid (mg)	385–1540	350–1400				
Linoleic/ALA	5–15.1		5–15		5–15	
ALA (mg)*	>55 (0.9% FA)	>50				
DHA (mg)*	12–30	11–27	ge21	ge16	ge18	ge16
AA (mg)	18–42	16–39	ge28	ge22	ge24	ge22
Carbohydrate (g)	11.6–13.2	10.5–12	9–20	6.0–15.4	7–17	5.4–15.5
Lactose						
Oligomers						
Sodium (mg)	69–115	63–105	69–115	46–88	69–115	53–105
Potassium (mg)	66–132	60–120	78–117	52–90	78–117	60–108
Chloride (mg)	105–177	95–161	107–249	71–192	107–249	82–226
Calcium (mg)	120–140	110–130	100–220	67–169	100–220	77–200
Phosphate (mg)	60–90	55–80	60–140	40–108	60–140	46–127
Magnesium (mg)	8–15	7.5–13.6	7.9–15	5.3–11.5	7.9–15	6.1–13.6
Iron (mg)	2–3	1.8–2.7	2–4	1.33–3.08	2–4	1.5–3.6
Zinc (mg)	1.1–2.0	1.0–1.8	1.0–3.0	0.67–2.3	1.0–3.0	0.77–2.7
Copper (µg)	100–132	90–120	120–150	80–115	120–150	92–136
Selenium (µg)	5–10	4.5–9.0	1.3–4.5	0.9–3.5	1.3–4.5	1.0–4.1
Manganese (µg)	≤27.5	6.3–25	0.7–7.5	0.5–5.8	0.7–7.5	0.5–6.8
Iodine (µg)	11–55	10–50	10–60	6.7–46.2	10–60	7.7–54.5
Vitamin A (IU; 1 µg RE = 3.33 IU)	400–1000	360–740	700–1500	467–1154	700–1500	538–1364
Vitamin D (IU)	800–1000		150–400	100–308	150–400	115–364
Vitamin E (mg α-TE)	2.2–11	2–10	6–12†	4.0–9.2	6–12†	4.6–10.9
Vitamin K (µg)	4.4–28	4–25	8–10	5.3–7.7	8–10	6.2–9.1
Vitamin C (mg)	11–46	10–42	18–24	12.0–18.5	18–24	13.8–21.8
Thiamin (µg)	140–300	125–275	180–240	120–185	180–240	138–218
Riboflavin (µg)	200–400	180–365	250–360	167–277	250–360	192–327
Pyridoxine (µg)	45–300	41–273	150–210	100–162	150–210	115–191
Niacin (µg)	380–5500	345–5000	3.6–4.8	2.4–3.7	3.6–4.8	2.8–4.4
B_{12} (µg)	0.1–0.77	0.08–0.7	0.3	0.2–0.23	0.3	0.23–0.27
Folate (µg)	35–100	32–90	25–50	17–38	25–50	19–45
Taurine (mg)			4.5–9.0	3.0–6.9	4.5–9.0	3.5–8.2
Inositol (mg)	4.4–53	4–48	32–81	21–62	32–81	25–74
Choline (mg)	8–55	7–50	14.4–28	9.6–21.5	14.4–28	11.1–25.5

*Ratio of AA to DHA should be in the range of 1.0–2.0 to 1 (wt/wt) and eicosapentaenoic acid supply should not exceed 30% of DHA supply.
†Maximum = 25 IU.
ESPGHAN, European Society of Paediatric Gastroenterology and Nutrition (Agostoni 2010); Tsang ELBW, extremely low birthweight (Tsang et al. 2005); VLBW, very low birthweight; MCT, medium-chain triglyceride; ALA, alpha-linolenic acid; FA, fatty acid; DHA, docosahexaenoic acid; AA, arachidonic acid; RE, retinol equivalent; α-TE, α-tocopherol equivalent.

Modified Bell's Staging in NEC

Table 20.20 Modified Bell's staging for necrotising enterocolitis (Bell et al. 1978; Walsh and Kliegman 1986)

STAGE	CLINICAL FINDINGS	RADIOGRAPHIC FINDINGS	GASTROINTESTINAL FINDINGS
I: Suspected	Apnoea and bradycardia, temperature instability, lethargy	Normal or mild ileus	Mild abdominal distension, increased gastric residuals
IIA: Confirmed Features of stage I plus:	Apnoea and bradycardia, temperature instability, lethargy	Ileus pattern with one or more dilated loops, bowel wall oedema, focal pneumatosis	Bloody stools, prominent abdominal distension, absent bowel sounds, mild abdominal tenderness
IIB: Confirmed severe Features of stage IIA plus:	Thrombocytopenia and mild metabolic acidosis	Any of: widespread pneumatosis, ascites or portal venous gas	Abdominal wall oedema, palpable loops, tenderness, sometimes cellulitis
IIIA: Advanced Features of stage IIB plus:	Mixed acidosis, oliguria, hypotension, coagulopathy, disseminated intravascular coagulation	Prominent loops, worsening ascites, no free air	Signs of peritonitis: marked tenderness and distension. Abdominal wall oedema and induration
IIIB: Advanced plus perforation Features of stage IIIA plus:	Shock, deterioration in laboratory values and vital signs	Pneumoperitoneum	Perforated bowel

Sizes of catheters used in Newborn

	Preterm	**Term**
Suction catheter (F)	8-10	10-12
Chest tube (F)	8-10	10-12
Orogastric tube (F)	5-6	6
Laryngoscope blade (F)	0	1

Drugs and dosages

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Department of Neonatology, Aster MIMS Kottakkal.

ACYCLOVIR:-

Dosage : 20 mg/kg/dose Q 8 hourly

Administration:

- Reconstitute 500 mg in 10 mg sterile water for injection
- Reconstituted solution is stable in room temperature for 12 hours
- Do not refrigerate
- Infusion solution concentration should be no greater than 7 mg/ml
- Solution is compatible with D5W and NS

ADENOSINE

Dose:

- Initial dose of 50 microgram/kg. Increase dose in 50 mic/kg increments in every 2 mt until sinus rhythm returns Usual maximum dose is 250 mic/kg

Administration

- As rapid IV push in 1-2 seconds.
- Infuse as close to IV site as possible
- Flush IV with saline immediately
- Compatible with D5W and NS

Formulation:

- Available in 2 ml vial containing 6mg adenosine dissolved in NS
- Do not refrigerate
- Store in room temperature

ADRENALINE

Formulation:

- 1in 1000 ampoule. Always use a 1:10000 (100mic/ml) solution

Dose:

- Resuscitation and severe bradycardia: 0.1 -0.3 ml/kg IV or 0.3 to 1 ml/kg endotra-trachial

IV infusion:

- 0.1 to 1 mic/kg/mt, titrated based on response

Administration:

- First dilute 1 ml adrenaline in 9 ml NS to make a 1 in 10000 solution
- Dilute 6 ml (600 micg)/kg to 50 ml with D5W or NS to make a 12 mcg/kg/ml solution.
- I ml/hour = 0.2 micg/kg/mt

AMIKACIN

PMA (weeks)	Post natal (days)	Dose (mg/kg)	Interval (hours)
≤29 *	0 - 7	18	48
	8 - 28	15	36
	≥29	15	24
30 - 34	0 - 7	18	36
	≥8	15	24
≥35	All	15	24
*Or significant asphyxia, PDA ortreatment with indomethacin			

Administration:

- Solution compatibility with D5W, D10W, D20W and NS
- For IV use dilute with a compatible solution to a concentration of 5 mg/ ml

AMPHOTERICIN B:-

Dosage

- 1-1.5 mg/kg every 24 hours

Administration

- 1-1.5 mg/kg
- IV infusion over 2-6 hours
- Reconstitute using D5W or preservative free sterile water to a concentration of 5mg/ml, , then dilute further using D5W to a concentration not greater than 0.1 mg/ml for infusion.
- Reconstituted solution is stable for 24 hours in room temperature or 7 days in refrigerator
- Do not flush IV or mix with NS
- Protect from light
- Compatible with all concentrations of dextrose

AMPHOTERICIN B LIPID COMPLEX:-

Dosage

- 5mg/kg per dose Q 24 hourly as infusion over 2 hours
- Dilute with D5W to 1-2 mg per ml dilution
- Do not freeze
- Protect from light
- Do not flush IV or mix with NS

AMINOPHYLLINE:

Dose:

- Loading dose: 8mg/kg IV infusion over 30 minutes or orally.
- Maintenance:1.5 to 3mg/kg/dose orally , or IV slow push every 8-12 hours after the loading dose .

In preterm infants, changing from IV aminophylline to oral theophylline requires no dose adjustment.

Available as aminophylline for IV use (25mg/ml) in 10 and 20 ml vials.

Solution compatibility: D5W, D10W and NS.

AMIODARONE

Dose and administration:

- Loading dose of 5 mg/kg over 30 to 60 mt
- Maintenance infusion: 7-15 microgram/kg/mt (10-20 mg/kg per 24 hours)
- Begin at 7 mic/kg/mt and titrate dose depending on response
- For prolonged infusions, the IV concentration should not exceed 2 mg/ml unless using a central line
- Consider switching to oral therapy within 24 to 48 hours
- Oral dose: 5-10 mg per kg per dose Q 12 hourly
- Compatible with D5W and NS

CAFFEINE CITRATE:

Dose

- Loading dose:20-25mg/kg of caffeine citrate IV over 30minutes or orally.
- Maintenance dose: 5-10 mg/kg /dose of caffeine citrate IV slow push or orally every 24 hours.
- Maintenance dose should be started 24 hours after the loading dose.
- Available as oral and solution each mL contains 20mg of caffeine citrate

CALCIUM - ORAL

Dosage

- 20-80 MG/KG elemental calcium per day orally in divided doses.

CALCIUM GLUCONATE

Dosage

- 10% IV FORMULATIONS (9.3 MG/ML elemental calcium): 2-8ml/kg/day.

CALCIUM CHLORIDE 10%

Symptomatic hypocalcemia-acute

Treatment:

35 -70 mg/kg/dose (0.35 - 0.7 ml/kg per dose, equiva-

lent to 10-20 mg/kg elemental calcium).

Dilute in appropriate fluid,then infuse in IV over 10-30minutes while monitoring for bradycardia. Stop infusion if heart rate is <100 beats/min.

DO NOT GIVE INTRA-ARTERIALLY

Maintenance treatment - 75-300mg/kg/day (0.75 to 3 ml/kg/day, equivalent to 20-80mg/kg elemental calcium). Administer by continuous IV infusion. Treat for 3-5 days, and follow serum concentrations periodically.

Exchange transfusion:

33mg/100ml citrated blood exchanged. Infuse IV over 10-30 minutes.

Calcium chloride 10%injection yields 27mg/ml elemental calcium (1.36 mEq/ml).

Solution compatibility:

D5W,D10W and NS.

CALCIUM GLUCONATE10%:-

Dosage

Symptomatic hypocalcemia-acute treatment :

100 to 200 mg/kg per dose (1 to 2ml/kg/dose, equivalent to 10 to 20mg/kg/dose, equivalent to 10 to 20 mg/kg elemental calcium)0

Dilute in appropriate fluid,then infuse in IV over 10-30 minutes while monitoring for bradycardia

DO NOT GIVE INTRA-ARTERIALLY.

Maintenance treatment - 200 -800mg/kg /day (2 to 8 ml/kg/day, equivalent to 20-80mg/kg elemental calcium). Administer by continuous IV infusion. Treat for 3-5 days, and follow serum concentrations periodically.

Exchange transfusion:

100 mg/100mL citrated blood exchanged. Infuse IV over 10 -30 minutes.

Calcium gluconate 10% injection yields 9.3 mg/ml elemental calcium (0.46 mEq/mL).

Solution compatibility:

D5W, D10W and NS.

CEFOTAXIME:-

Dosage

50 mg/kg/dose IV infusion over 30 minutes

PMA (weeks)	Post natal (days)	Dose (mg/kg)	Interval (hours)
≤29 *	0 - 7	18	48
	8 - 28	15	36
	≥29	15	24
30 - 34	0 - 7	18	36
	≥8	15	24
≥35	All	15	24
*Or significant asphyxia, PDA ortreatment with indomethacin			

Formulation :

- Iv is available as 150 mg/ml solution.
- Dilute using D5W, NS or RL to a maximum concentration of 18mg/ml
- Maximum infusion rate 30 mg/minute
- Compatible with D5W, D10W and NS.

COLISTIN:

Dose:

- 5mg per kg per day, or 50,000 to 75,000 IU/kg/day. In divided doses Q 8 hourly

Formulation:

- colistimethete sodium in vials of 150 mg colistin base

Administration:

- Reconstitute with 2 ml of WFI toyeild 75mg/ml base. Gently swirl to avoid frothing
- Administer as sow IV over 3-5 minutes

DEXAMETHASONE

DOSE

- DART trial protocol: 0.075mg/kg/dose every 12 hours for 3 days, 0.05mg/kg/dose every 12 hours for 3 days, 0.025mg/kg/dose every 12 hours for 2 days and 0.01mg/kg/dose every 12 hours for 2 days. Doses may be administered IV slow push or orally.
- Available as 4mg/ml and 10mg/ml.

Solution compatibility:

- D5W, D10W and NS.

DIAZOXIDE:

Dosage:

- 2 - 5 mg/kg/dose orally given every 8 hours. Begin therapy at the higher dosage and taper by response.
- Available as an oral suspension, 50mg/ml concentration and tablet 50 mg

DIGOXIN

Dosage

- Loading dose: Digitalization is only done when treating arrhythmias or acute CCF. Give over 24 hours in 3 divided doses, as IV slow push over 5 - 10 minutes.
- Oral dose should be 25% greater than IV
- Do not give as IM

Total loading dose		
PMA (weeks)	IV (mcg/kg)	PO (mcgkg)
≤29	15	20
30-36	20	25
37-48	30	40
≥49	40	50
Divide in to three doses over 24 hours		

Maintenance dose			
PMA (weeks)	IV (mcg/kg)	PO (mcgkg)	Interval (hours)
≤29	4	5	24
30-36	5	6	24
37-48	4	5	12
≥49	5	6	12
Titrate based on clinical response			

Formulations:

- Injection 100 mcg/ml and elixir 50 mcg/ml
- Compatible with D10W, NS, sterile water for injection
- Store at room temperature

DOBUTAMINE

Dose:

- 5-25 mic/kg/mt, titrated based on clinical response

Formulation:

- 250mg vial. Store below 25 degree C

Administration:

- Add 10 ml of WFI to a 250 mg vial to make1 25mg/ml solution
- Dilute 1.2 ml (30mg)/kg to 50 ml of D5W or NS to make a 30 mg/kg/50 ml solution (max 5mg/ml)
- 1 ml/hour of this solution will give adose of 10 mcg/kg/mt
- Use syringe pump for infusion
- Stable in parenteral solutions for 24 hours
- Don't give in same line as heparin, NaHCO3 or penicillin

DOMPERIDONE

Dose:

- 0.3mg/kg/dose Q 8-12 hourly orally

Presentation:

- 1ml/ml syp and 10 mg/ml drops

DOPAMINE

Dose:

- 2-20 mcg/kg/mt, titrated based on clinical response

Formulation:

- 200mg/5ml ampoule. Store below 25 degree C

Administration

- Dilute 0.75 ml (30mg)/kg to 50 ml of D5W or NS to make a 30 mg/kg/50 ml solution (max 3.2mg/ml)
- 1 ml/hour of this solution will give a dose of 10 mcg/kg/mt
- Use syringe pump for infusion
- Stable in parenteral solutions for 24 hours

ERYTHROMYCIN

For treatment of feed intolerance due to dysmotility: 10mg/kg/dose per orally Q 6 hourly for 2 days, followed by 4mgkg/dose orally Q 6 hourly for 5 days

Available as 200mg and 400 mg per 5 ml suspension and 1ml/100mg drops

FENTANYL

Sedation and analgesia:

- Bolus dose: 0.5 to 4 mcg/kg per dose IV slow push repeated as required
- Infusion: 1-5 mcg/kg/hour
- Compatible with D5W, D10W or NS

FERROUS SULFATE

Dosage:

- 2mg/kg/day of elemental iron for growing premature infants.(maximum of 15mg/day).
- Begin therapy after 2 weeks of age.
- Infants with birth-weights <1000grams may need 4mg/kg/day.
- 6mg/kg/day of elemental iron for patients receiving erythropoietin. Administer orally in 1 or 2 divided doses, preferably diluted in formula.

FLUCONAZOLE

Dosage:-

- Invasive candidiasis: 12-25mg/kg loading dose, then 6-12 mg/kg per dose IV infusion over 30 mt or orally
- Consider higher dose for candida strains with MIC of >4mic/ml

- Consider extended dosing interval if serum creatinine is >1.3
- Prophylaxis: 3mg/kg/dose twice weekly as IV infusion or orally
- Oral thrush: 6mg/kg on day 1, then 3 mg/kg/dose Q 24 hourly orally

Gest age (weeks)	Post natal (days)	Interval (hours)
≥29	1-14	48
	>14	24
30 and older	0-7	48
	>7	24

Formulations:

- As premixed solution for IV injection 200mg/100ml or 400 mg/200 ml
- Tablets of 50, 150 mg 200 mg

Administration:

- Store in room temperature
- Do not freeze
- Compatible with D5W and D10 W

GENTAMICIN:-

- IV infusion over 30 mt
- IM injection has variable absorption

PMA (weeks)	Postnatal (days)	Dose (mg/kg)	Interval (hours)
≤29*	0-7	5	48
	8-28	4	36
	?29	4	24
30-34	0-7	4.5	36
	?8	4	24
≥35	All	4	24
*or significant asphyxia, PDA, treatment with indomethacin			

Administration:

- Compatible with D5W, D10W, NS

GLUCAGON

Dosage

- 200 mcg/kg/dose (0.2mg/kg/dose) IV push, IM or subQ.
- Maximum dose: 1mg.
- Continuous infusion: Begin with 10 - 20 mcg/kg/hour (0.5 to 1 mg/day)
- Available as 1mg single dose vials.

HYDROCORTISONE

Dosage:

- Physiologic replacement: 7-9mg/m2 /day IV or orally, in 2 or 3 doses.
- Treatment of pressor and volume resistant hypo-tension: 20-30mg/m2 /day IV in 2 or 3 doses, or approximately 1mg/kg/dose every 8 hours.
- Treatment of chorioamnionitis exposed ELBW infants to decrease risk of CLD:
- Initial dose : 0.5mg/kg/dose IV every 12 hours for 12 days, followed by 0.25mg/kg IV every 12 hours for 3 days.

Weight (kg)	Surface area (sq meters)
0.6	0.08
1	0.1
1.4	0.12
2	0.15
3	0.2
4	0.25
BSA(m2)=(0.05xkg)+0.05	

- Hydrocortisone sodium succinate is available as powder for injection in 2 mL vials containing 100mg.
- Solution compatibility: D5W,D10W and NS.

IBUPROFEN LYSINE

- First dose:10mg/kg.
- Second & third dose: 5mg/kg.
- Administer IV by syringe pump over 15minutes at 24 hour intervals.
- Contraindicated in preterm neonates with 1) infection 2) active bleeding 3) thrombocytopenia or coagulation defects, 4) NEC 5) significant renal dysfunctions 6) CHD with ductal dependent systemic blood flow.
- Available as 10mg/ml sterile solution for injection in 2ml single use vials.
- Solution compatibility : NS and D5W.

INDOMETHACIN

Dosage:

- Short course: 0.2 mg/kg/dose Q 12 hourly for 3 doses
- Long course: 0.1mg/kg/dose Q 24 hourly for 6 doses
- Formulation: 25 mg capsule and 1 mg vial for IV

Administration

- IV infusion over 20 to 30 mt
- Store below 25 degree C
- Protect from light

IMIPENEM

Dose

20-25 mg per kg per dose Q 2 hourly as infusion over minimum 30 minutes. Works better as infusion over 2-3 hours.

Formulations

As powder for injection in 250 mg and 500 mg vials

Administration

- Reconstitute with 10 ml of compatible diluents, solution is stable for 4 hours in room temperature and 24 hours when refrigerated.
- Maximum concentration for infusion is 5mg/ml
- Compatible with D5W, D10W, NS.
- IVIG (HUMAN)

Dose

- 500 - 1000 mg per kg per dose over 2-6 hours
- Most studies have used single dose, although additional doses can be used at a interval of 24 hours

Administration

- As slow IV
- Do not mix IVIG products from different manufacturers
- Do no freeze

LEVOTHYROXINE

Dosage

- Initial oral dose-10-14 mcg/kg /dose orally every 24 hours.
- Initial IV dose -5-8 mcg/kg/dose every 24 hours.
- Available as scored tablets ranging from 25 -300 mcg/ tablet.

IRON DEXTRAN

Dosage

- 0.4 - 1mg/kg (400 -1000mcg/kg)/day IV continuous infusion in Dex/AA solutions containing at least 2% amino-acids.
- Available as a 50mg/ml concenterations in 2ml single dose vials.

INSULIN:-

Dosage:

- Hyperglycemia: Continuous IV infusion:0.01 to 0.1 unit/kg/hour.
- Intermittent dose: 0.1 - 0.2 unit/kg every 6 to 12 hours subQ.
- Hyperkalemia: Initial -Regular insulin 0.1 to 0.2 unit/kg/hour in combination with 0.5g/kg/hour of dextrose given as continuous IV infusion.
- Regular human insulin is available as 100 units/ml concentration in 10mL vials. For subcutaneous administration, dilute with sterile water or NS to a concentration of 0.5 or 1unit/ml.
- Keep refrigerated.
- Solution compatibility: D5W, and D10W and NS.

LEVOTHYROXINE

Dosage:

- Initial oral dose: 10-14 mcg/kg/dose orally every 24 hours. (37.5 to 50 mcg/dose for an average term infant.) Dosage is adjusted in 12.5 mcg increments. Always round upward.
- Initial IV dose: 5 to 8 mcg/kg/dose every 24 hours.
- Available as scored tablets ranging from 25 - 300 mcg/ tablet.

LINEZOLID

Dose

- 10mg/kg/dose Q 8 hourly as infusion over 30-120 mt
- For preterm babies who are <7 days of age, the interval is Q 12 hourly
- Oral dose is same as IV

Formulation:

- Available as 2 mg/ ml in single use, ready to use 1000ml, 200ml and 300 ml infusion bags
- Oral preparation as 00 mg per 5 ml

Administration

- Store in room temperature
- Do not freeze
- Compatible with D5W, NS and RL

MAGNESIUM SULFATE

Dosage

- Resuscitation: 25-50mg/kg iv/intraosseous rapid infusion.
- Hypomagnesemia: 25-50mg/kg iv infusion over 30-60 minutes; repeat dose as necessary.
- Daily maintenance requirements: 0.25 - 0.5 meq/kg/day iv.
- Administration: Must be diluted prior to IV administration.
- Available as 50% concentration in 2-,10- and 50-ml single dose vials containing 500mg/ml of magnesium sulfate which provides 4.06 mEq each of magnesium and sulfate.
- Solution compatibility:D5W, NS,LR and Dex/AA solutions.

MEROPENEM

Dose

- Sepsis: 20 mg / kg Q 8 hourly
- <32 weeks: For ≤14 days Q 12 hourly and for >14 days Q8 hourly
- ≥32 weeks: for ≤7 days 12 hourly and for >14 days Q 8 hourly

- Meningitis and pseudomonas sepsis 40 mg per kg per dose in all ages
- Give as IV infusion over 30 mt. Longer infusions up to 4 hours has better efficacy

Formulations:

as powder for injection of 125, 250, 500 mg and 1g

Administration:

- Solution reconstituted with water for injection is stable for 2 hours in room temperature and for 12 hours when refrigerated
- Solution prepared in sterile water for injection or NS to a concentration of 1-20 mg per ml is stable in plastic syringes for up to 48 hours when refrigerated
- Solutions prepared in D5w are stable for lesser durations
- Compatible with D5W, D10W and NS

METOCLOPRAMIDE

Dosage:

- 0.033-0.1MG/KG/DOSE orally or IV slow push every 8 hours.
- Metoclopramide can cause tardive dyskinesia. The risk increases with duration of treatment and total cumulative dose.
- Available as 5mg/ml injectable solution.
- Protect from light.
- Solution compatibility: D5W and NS.

METRONIDAZOLE:-

Dose

- Loading dose: 15 mg per kg orally or as IV infusion over 2 hours
- Maintenance dose: 7.5 mg per kg orally or as IV infusion over 2 hours. Begin one dosing interval after loading dose

PMA (weeks)	Post natal (days)	Interval (hours)
≤29	0-28	48
	>28	24
30-36	0-14	24
	>14	12
37-44	0-7	23
	>7	12
≥45	All	8

Formulation:

- as 5ml/ml solution in ready to use , single use 100 mg plastic bottle
- Protect from light
- Do not refrigerate
- Store in controlled room temperature
- Compatible with D5W and NS

MILRINONE

Dose:

- Loading dose of 50micg/kg given IV over 15-30 minutes. Loading dose can be reduced to 25 micg/kg or omitted in patients with hypotension

Formulation:

- 1 mg/ml concentration in 10, 20 and 50 ml vials

Administration:

- The loading dose can be given as diluted in 10-20 ml or undiluted
- For infusion, dilute to a concentration of 200 to 400 micg/ml with D5W, RL or NS

MORPHINE

Dosage

- 0.05 - 0.2Mg/kg/dose IV at-least 5 minutes, IM or subcutaneous
- Continuous infusion: Loading dose of 0.1 - 0.15 mg/kg over 1 hour followed by 0.01 -0.02 mg/kg/hour.
- Initial treatment of neonatal abstinence syndrome:0.03 -0.1 mg/kg/dose orally every 3-4 hours. Wean dose by 10% - 20% every 2-3 days based on abstinence scoring.
- Injectable solutions are available in dosage ranging from 0.5 - 50mg/ml.
- Oral morphine Sulfate solutions available in concentrations of 2,4 and alcohol free 20mg/ml.
- Solution compatibility: D5W, D10W and NS.

NEOSTIGMINE

Dosage

- Mysthania gravis:0.1mg IM (give 30 minutes before feeding).1 mg orally (give 2 hours before feeding). Dose may have to be increased and should be titrated.
- Reversal of neuromuscular blockade: 0.04 - 0.08 mg/kg IV, in addition to atropine 0.02mg/kg.

OCTREOTIDE

Dosage

Treatment of hyperinsulinemic hypoglycemia:

Initial dose:1 mcg/kg/dose every 6 hours subQ or IV.

Maximum dose: 10mcg/kg/dose every 6 hours.

Treatment of Chylothorax:

Begin at 1mcg/kg/hour IV continuous infusion.

Maximum dose:10mcg/kg/hour

OMEPREZOLE

Dosage

- 0.5- 1.5MG/KG/DOSE orally, once a day.
- Available as 20mg powder for suspension packet.

PARACETAMOL

Dose

For analgesia:

- Oral loading dose of 20-25 mg/kg and maintenance dose of 12-15 mg/kg/dose
- Rectal loading dose of 30mg/kg and maintanence of 12-18 mg/kg/dose

Dose interval:

- Q 6 hourly in term babies. For preterm babies ?32 weeks PMA interval is Q 8 hourly and for <32 weeks Q 12 hourly

For PDA treatment:

- 15 mg per kg per dose Q 6 hourly (oral or IV) for 3 days. If the PDA not closing by day 3, the treatment can be extended to 6 days

PENICILLIN G (BENZYLPENICILLIN)

- Meningitis: 75,000 to 100,000 U per kg per dose as IV infusion over 30 mt or IM
- Bacteremia: 25,000 to 50,000 U per kg per dose as IV infusion over 15 mt or IM
- Congenital syphilis: 50,000 U per kg per dose as IV infusion over 15 mt, given every 12 hourly in first 7 days of life, and every 8 hourly there after, irrespective of GA. Total duration is 10 days.

PMA (weeks)	Post natal (days)	Interval (hours)
≤29	0-28	12
	>28	8
30-36	0-14	12
	>14	8
37-44	0-7	12
	>7	8
≥45	All	6

- Formulation: Available as penicillin G sodium and penicillin G potassium.

Administration

- Reconstitute the 5 million unit vial with 8 ml sterile water for injection to make aa final concentration of 500,000 u per ml. A 100,000 u per ml solution can be made by adding 10 ml of reconstituted solution to 40 ml sterile water for injection
- Reconstituted solution is stable for 7 days when refrigerated. Penicillin G sodium solution is stable for 3 days after reconstitution.
- 1 million unit is the equivalent of 600 mg
- Compatible with D5W, D10W and NS

PENICILLIN G BENZATHINE

- Congenital syphilis: 50,000 unit/kg IM single dose

Administration

- For IM injection only
- Available in a concentration of 600,000 u per ml
- Store in fridge, donot freeze

PENICILLIN G PROCAINE

- Congenital syphilis: 50,000 u/kg/dose IM once daily for 10 days

Administration

- For IM use only
- Available in a concentration of 600,000 u per ml
- Store in fridge, donot freeze

PIPERACILLIN:-

Dosage

- 50-100MG/KG/DOSE IV infusion by syringe pump over 30 minutes or IM.

PMA (weeks)	Post natal (days)	Interval (hours)
≤29	0-28	12
	>28	8
30-36	0-14	12
	>14	8
37-44	0-7	12
	>7	8
≥45	ALL	6

- Available as powder for injection in 2g, 3g, 4g and 40g vials.
- Reconstitute sterile water to make final concentration of 200mg/ml.
- Reconstituted solution stable for 24 hours at room temperature, 2 days refrigerated.
- Solution compatibility: D5W, D10W and NS.

PIPERACILLIN-TAZOBACTAM

Dose

- 50-100 mg (of pioeracillin) per kg per dose IV infusion over minimum 30 minute.

PMA (weeks)	Post natal (days)	Interval (hours)
≤29	0-28	12
	>28	8
30-36	0-14	12
	>14	8
37-44	0-7	12
	>7	8
≥45	ALL	6

- Formulation: As 2, 25, 3.75 and 4.5 g powder for injection

Administration

- Reconstitute with sterile water to make a solution of 200 mg per ml (of piperacillin)
- Reconstituted solution is stable for 24 hours in room temperature and for 2 days when refrigerated
- Use 400 mg per ml concentration for IM use
- Compatible with D5W, D10W, RL and NS

POTASSIUM CHLORIDE:-

Dosage

- Initial oral replacement therapy:0.5-1 mEq/kg/day divided and administered with feedings.
- Acute treatment of symptomatic hypokalemia:Begin with 0.5 -1 mEq/kg IV over 1 hour, then reassess. Maximum concentration:40 mEq/L for peripheral , 80 mEq/L for central venous infusions.
- Available as 2 mEq/ml solution.
- Always dilute before administration.

PROSTAGLANDIN E1 (ALPROSTADIL)

Dose:

- Initial dose: 0.05 to 0.1microgram/kg/mt as continuous IV infusion
- Maintenance dose: as low as 0.01 mic/kg/mt. titrate the dose depending on the clinical response (Saturation/ BP/ adverse effects)
- Higher doses may be required in left sided obstructive lesions
- Sample infusion: Mix 1 ampule (500 microgram) in 49 ml of compatible solution (D5W, NS) yielding a solution of 10 mic/ml. Infuse at 0.6 ml per kg per hour to provide a dose of 0.1 mic/kg/mt

PYRIDOXINE

Dosage :

- Initial diagnostic dose: 50-100mg IV push, or IM.
- Maintenance dose: 50-100mg orally every 24 hours. High doses may be required during periods of inter-current illness.
- Injectable form available in concentration of 100mg/ml.
- Protect from light.

RANITIDINE

Dosage :

- Oral-2mg/kg/dose every 8 hours.
- IV:TERM-1.5 mg/kg/dose every 8 hours slow push.
- Pre term-0.5mg/kg /dose every 12 hours slow push.
- Continuous IV infusion-0.0625mg/kg/hour; dose range 0.04-0.1 mg/kg/hour.
- Available as a 1mg/ml preservative free solution for injection in 50ml single-dose plastic container and a 25mg/ml injectable solution in 2 and 6 ml vials.
- Oral solution available as 15mg/ml
- Also available as 150 & 300mg tablets.
- Solution compatibility:D5W ,D10W and NS.

SILDENAFIL

Dosage

- IV: Administer a loading dose of 0.4mg/kg over 3 hours, followed by continuous infusion of 1.6mg/kg/day.
- ORAL: 0.5 to 2mg/kg/dose every 6 to 12 hours.
- Available as 20mg tablets and as a single use vial containing 10mg of sildenafil equivalent to 0.8mg sildenafil per ml.

SODIUM NITROPRUSSIDE

Dosage

- 0.25 - 0.5 mcg/kg/min continuous IV infusion by syringe pump. Usual maintenance dose is <2 mcg/kg/min. For hypertensive crisis, may use up-to 10mcg/kg/min, but for no longer than 10 minutes.
- Available as powder for injection in 2 mL single dose 50mg vials.
- Do not administer reconstituted drug directly from vial.
- Solution compatibility: D5W, NS AND LR only.

SODIUM BICARBONATE

Dosage

- Dosage based on base deficit: hco3 needed (meq)= hco3 deficit (meq/l) x(0.3 x body wt(kg))administer half of calculated dose, then assess need for remainder.
- Usual dosage: 1 to 2 mEq/kg/ IV over at-least 30 minutes.
- Recommended dilution: 0.25 mEq/mL.
- Maximum concentration: 0.5 Eq/ml.

- Can be administered by continuous IV infusion or orally.
- Available as 4% (0.48 mEq/ml), 4.2% (0.5 mEq/ml), 5% (0.6 mEq/ml), 7.5% (0.9 mEq/mL) and 8.4% (1 mEq/mL). Maximum concentration used in neonates is 4.2%.
- Solution compatibility: D5W, D10W and NS.

SPIRONOLACTONE

Dosage:

- 1-3 mg/kg /dose every 24hours.
- Available in 25mg, 50mg and 100mg tablets.
- Suspensions are stable for 1 month refrigerated.

VANCOMYCIN

- Bacteremia: 10mg/kg/dose
- Meningitis: 15mg/kg/dose
- As IV infusion over 2 hours

PMA (weeks)	Post natal (days)	Interval (hours)
≤29	0-14	18
	>14	12
30-36	0-14	12
	>14	8
37-44	0-7	12
	>7	8
≥45	All	6

- Formulation Available as powder for injection in 250,500 mg and 1 g vials

Administration

- Reconstitute with sterile water to make a solution of 50 mg/ml
- Reconstituted solution is stable for 4 days when refrigerated
- Dilute prior to administration with NS or D5W to a maximum concentration of 5 mg/ml
- Compatible with D5W, D10W and NS

SURFACTANT

(Natural, animal derived)

CUROSURF

Dosage

- Initial dose: 2.5 ml/kg /dose intratracheally, divided into 2 aliquots followed by up to two subsequent doses of 1.25 ml/kg/dose administered at 12 hour interval if needed.
- Clear the trachea of secretions. Shorten a 5F end hole catheter so the lip of the catheter will protrude just beyond end of ET tube above infant's Carina.
- Available in 1.5 mL and 3 mL vials.

NEOSURF

Dose

- 5 ml / kg
- Interval between doses is 12 hours for maximum 2 doses

SURVANTA

Dosage

- Initial dose: 4 ml/kg /dose intratracheally, divided into 4 aliquots.
- Prophylaxsis: first dose is given as soon as possible after birth, with up to 3 additional doses in the first 48 hours of life, if indicated.
- Rescue treatment of RDS:Up to 4 doses in first 48hours of life, no more frequently than every 6 hours.
- Before administration, allow to stand at room temperature for 20minutes, or warm in the hand for at least 8 minutes.
- Artificial warming methods should not be used.
- Available in 4 and 8 mL single use vials.

VITAMIN A

Dosage

- Parenteral treatment of vitamin a deficiency: 5000 units im 3 times weekly for 4 weeks.
- Do not administer IV
- Available as Aquasol A Parenteral 50000 units per ml, equivalent to 15mg retinol per ml, in 2ml vials.
- Protect from light. Do not freeze.

VITAMIN D

Dosage

- 400 units per day orally.
- Treatment of vitamin D deficiency:1000 units/day orally.
- Vitamin D supplements are available as vitamin D2 (ergocalciferol) and vitamin D3 (Cholecalciferol)

VITAMIN E

Dosage

- 5-25 UNITS /DAY orally. Dilute with feeding. Do not administer simultaneously with iron-iron absorption is impaired.
- Available as liquid drops:Aquavil E, 15units per 0.3ml.

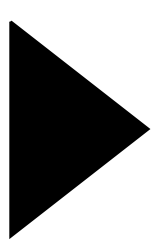
VITAMIN K1

Dosage

- Recommended prophylaxis:0.5 to 1 mg IM at birth.
- Preterm infants less than 32 weeks of gestation:
- B.WT->1000GRMS: 0.5mg IM.
- B.WT-<1000 GRAMS :0.3mg/kg IM.
- Available as a 2 mg/ml aqueous dispersion in 0.5ml ampules and 10mg/ml aqueous dispersion in 1mL ampules and 2.5 and 5 ml vials.
- Solution compatibility:D5W, D10W and NS.

Abstracts

of Thrissur Neocon 2017



Hypoglycemia: Some Bittersweet Facts

Dr. Preetha Remesh MD., MRCP., MRCPCH

HODNeonatology,AsterMIMSCalicut

- Where does the brunt of Hypoglycemia Induced brain Injury fall?

  Neurons of Cerebral cortex, Hippocampus & Caudate nucleus

- How does this Injury come about?

  Selective neuronal necrosis by apoptosis following mitochondrial membrane & DNA damage and excitotoxic damage mediated by the elevated levels of glutamate via NMDA receptor stimulation.

- Blood sugar at birth is ~ 70% of maternal value.

  This gradient was, incidentally, necessary in the fetal life to ensure a continuous flow of glucose from mother.This relatively low blood sugar does help to kick start important physiological processes vital for sustaining life i.e., Gluconeogenesis & Glycogenolysis.

- Whipple's Triad: -

  Low Blood sugar value + Clinical signs+ complete resolution once normoglycemia is achieved.

- Asymptomatic Hypoglycemia:-

  This is when compensatory mechanisms have succeeded in maintaining the mileau interior intact; but at some cost. Glycogen reserves have been eroded into, leaving baby vulnerable to future energy challenges. Thus, asymptomatic hypoglycemia is not entirely inconsequential. Another noteworthy point is, for each symptomatic hypoglycemia we pick up, there has been quite a while of asymptomatic hypoglycemia. BUT, it must be noted that asymptomatic hypoglycemia cause no neuro developemental sequelae.

- Signs & Symptoms:-

  1. Systemic manifestations of Glucopenia:- Poor feeding, Irritability, hypothermia, apnea, tachycardia, tachypnea
  2. Manifestations of Neuroglucopenia:- Tremors, changes in level of consciousness, seizures, coma.

- Operational Threshold: -

  This concept has replaced the futile quest for a magic numerical value above which all is well. OT is the value of blood sugar at which we have to sit up & act. This will mean actively ensuring frequent feeds & monitoring for any untoward signs. Thus OT is an indication for action & not a diagnosis.

  At < 24 hours, for a term healthy baby, OT is 30-35 mg%

  At < 24 hours, for a sick/preterm/IUGR baby, OT is 45mg%

- Onset of Hypoglycemia may even be prenatal!

  Falling or low estriol levels & hypoxia in mother both can affect the transplacental transport of glucose and therefore, the fetal hepatic stores.

- Neuro developemental outcome of the vulnerable group comprising of SGA, LGA,IUGR, IDM and late preterm babies is worse than that of a healthy term baby. In some cases, there is associated hypoglycemia too. BUT, no study has shown that preventing/treating hypoglycemia in this group results in a better outcome either.

- Inborn errors of metabolism do present as refractory hypoglycemia.

  But in actual fact, only a very few present primarily as this. Fatty Acid oxidation defect, one of the major disorders of gluconeogenesis is a striking example.(MCHAD/LCHAD). Others are Galactosemia,Glycogen Storage Disorder 1 & Glycogen debrancher enzyme deficiency Correction of hypoglycemia may not totally revive the babies, as there is the buildup of toxic metabolites to reckon with as well.

- Finally, HYPOGLYEMIA IS A LITOGEN! Just like a mitogen; a litogen is induces litigations!

Intrauterine Infections : Obstetrician's Perspectives

Mr. Ashok Kumar

MD, FRCOG, Consultant Obstetrician & Gynaecologist

Pregnant women are susceptible to getting various viral, bacterial and parasitic infections which can have harmful effects on babies and, cause increased perinatal morbidity and mortality.

The common causative organisms are toxoplasma gondi, rubella virus, cytomegalovirus, herpes, enterovirus, syphilis, chickenpox, human immunodeficiency virus and Parvovirus B19.

Congenital infections are usually acquired by trans-placental entry of the organism. Infection can occur during labour and delivery as well.

Infections acquired during pregnancy may result in fetal loss, intrauterine growth retardation, central nervous system damage and still birth. Post natally, congenitally acquired infections may present as microcephaly, hepatosplenomegaly with abnormal liver function tests, thrombocytopenia, mental retardation, behavioural problems, and physical abnormalities such as cerebral palsy. Some cases can be totally asymptomatic.

Each of the infecting agents can its own characteristic features

For example, maternal CMV infection is characterised by ocular defects, including chorioretinitis, microphthalmos, cataracts and optic atrophy; sensorineural deafness.

There is a review of the current approach to the pre-natal diagnosis and management of the common and most clinically relevant congenital infections: CMV, Parvovirus B19, toxoplasma gondi, rubella and varicella- zoster virus.

Relevant tests are usually done when there is symptomatic maternal infection, finding of sonographic markers of fetal infection during ultrasound infection and when there is a maternal exposure to the pathogen.

There are many laboratory tests available to detect these organisms. The interpretation of the results can be complex.

Clinicians should take consideration of clinical presentation, the timing of the test in relation to exposure to the agent, the benefits and limitation of pre-natal diagnosis and the effectiveness of potential treatment.

Care should be optimally provided by a multidisciplinary team involving obstetricians, virologists, fetal medicine specialists and neonatologists.

The approach to the pre-natal diagnosis of congenital infection varies according to the gestational age and the likely agent. The first step usually is to confirm maternal infection. This is most frequently done by testing pathogen specific Ig and IgM.

Amniocentesis to test for the presence of RNA or DNA by PCR is the mainstay of diagnosis of fetal infection in some cases but the timing of the test in relation to the likely point at which transmission occurred is crucial.

The detection of the virus alone does not mean that there is a fetal damage and a negative result does not completely exclude the possibility of fetal infection. Ultrasound surveillance is the most important way to determine the degree of damage, but it has limitations in accurately predicting the outcome of the baby.

Therapeutic options are limited. Intrauterine blood transfusion can be helpful in cases of anaemia due to parvovirus infection and maternal antibiotic treatment for toxoplasmosis infection.

Newer Modes of ventilation: HFOV with VG and NAVA

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HFOV with VG:

High-frequency oscillatory ventilation (HFOV) is characterized by an effective gas exchange using tidal volumes equal to or less than the dead space volume at supra-physiological frequencies. Carbon dioxide removal is mostly relate to the tidal volume generated during high-frequency ventilation, and this high-frequency expired tidal volume (VThf) is known to be close to the airway dead space. Also, the frequency has a role in CO2 removal and has an independent effect on the distribution of the gas within the airways. During HFOV, several mechanisms of gas exchange have been described, the combination of which is responsible for its ventilatory efficiency.

VThf delivery depends on the ventilator characteristics, and although VThf monitoring is useful in daily practice, most of the ventilators providing HFOV do not display or measure the VThf. However, in some new HFOV ventilators it is possible to adjust directly the VThf constant due to the volume guarantee (VG). VG is a well-documented volume target ventilation modality combined to synchronize conventional tidal ventilation. HFOV with VG enables to turns ventilator into a powerful high frequency oscillator with guaranteed VThf. This strategy will like to reduce both barotrauma and volutrauma.

NAVA:

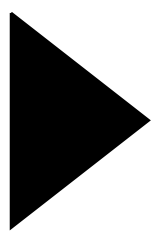
Neurally Adjusted Ventilatory Assist (NAVA) is a mode of ventilation where the individual patient's own respiratory drive (Edi, see above) controls timing and assist delivered by the ventilator. The electrical activity of the diaphragm (Edi) is a diagnostic tool that allows for continuous bedside monitoring of your patient's breathing effort. It is measured with the help of small sensors on the patient's feeding tube.

Personalized ventilation provides unique patient insight and ventilation capabilities. It consists of a diagnostic tool that helps you monitor diaphragm activity (Edi) on the ventilator screen and a ventilation mode (NAVA) that uses the diaphragm activity to deliver assist adapted to the patient. This personalized ventilation can help in reducing complications, increase patient synchrony - comfort, reducing need for sedation and ability to wean patients earlier.

During normal respiration, a spontaneous breath begins with an impulse generated by the respiratory centers in the brain. This impulse is then transmitted via phrenic nerves and electrically activates the diaphragm, leading to a muscle contraction. The diaphragm contracts into the abdominal cavity, which leads to a descending movement. This creates a negative alveolar pressure and an inflow of air. The signal that excites the diaphragm is proportional to the integrated output of the respiratory center in the brain and controls the depth and cycling of the breath.

With NAVA ventilation the electrical discharge of the diaphragm is captured by a special catheter fitted with an array of electrodes (the Edi catheter) and visualized on the ventilator screen. This is Edi, the electrical activity of the diaphragm. The Edi catheter is placed in the esophagus much like an ordinary feeding tube. With NAVA, Neurally Adjusted Ventilatory Assist, the Edi is used to deliver ventilation in time with and in proportion to the diaphragm activity.

Miscellanea



“All that wheezes is not asthma”

Dr PMC Nair

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Hon: Consultant, KIMS,TVM

Case report:

One fine morning the sun rose as usual in the East , with pleasant blue sky and birds chirping as usual and I was on duty. Around 10 am, a term male baby was brought to our NICU with respiratory distress soon after birth. On examination ,baby was term with a birth weight of 3.36 kg with a respiratory rate of 70/mt and suprasternal retractions. Clinical examination otherwise was unremarkable. Chest X-ray and blood gases were initially normal.ENT examination done was unremarkable. Echocardiography gave a suggestion of persistent pulmonary hypertension (PPHN) with open foramen ovale (PFO) and ductus arteriosus(PDA), but clinically baby had no signs of PPHN.

Child was straining with suprasternal retractions and was becoming sicker with occasional apnoeic episodes. Refusing to accept the diagnosis of PPHN, we went ahead with CT scan chest which also was inconclusive. Finally we took the baby for MR- cine angiography which clinched the diagnosis.

Discussion:

Two important things are highlighted by this case. First, we should not forget the basics. Any baby with supra-sternal retractions, think of the possibility of an upper airway obstruction. Secondly similar to the fact that “all that wheezes is not asthma” we should not conclude that any newborn with suprasternal retactions, noisy breathing or stridor is laryngotracheomalacia or Congenital laryngeal stridor(CLS). The differential diagnosis of respiratory distress with suprasternal retractions (upper airway obstruction) include choanal atresia, Pierre robin sequence, macroglossia, laryngeal web or cyst, laryngomalacia, trachea-esophageal fistula, vascular rings, external compression from a neck mass, vocal cord paralysis, hemangiomas or papillomas and subglottic stenosis

In this baby MR cine angiography revealed a right anomalous subclavian artery compressing the trachea and along with the patent ductus arteriosus was forming a constricting vascular ring around the trachea.(Fig:1)

On the operation table the right subclavian reanastomosed and PDA ligated. There was even an indentation on the trachea. The baby recovered slowly, weaned off the ventilator and discharged home after a week. On follow-up the baby is doing well, normal growth and development and no stridor or respiratory problems. Aberrant right subclavian artery (ARSA) also known as arteria lusoria is the commonest of the aortic arch anomalies with an estimated incidence of 0.5 -2% and a female preponderance. Instead of being the first branch (with

the right common carotid as the brachiocephalic artery), it arises on its own as the fourth branch, after the left subclavian artery and then hooks back to reach the right side. Its relationship to the oesophagus is variable: 80% posterior to esophagus and 5% anterior to trachea as in this case.

Clinical presentation is often asymptomatic but around 10% may have tracheo-oesophageal symptoms and dysphagia (dysphagia lusoria) .Arteria lusoria, was first described by Bayford in 1794 in a 62-year-old woman who died after years of dysphagia. Adults typically present with symptoms of dysphagia; infants more often present with respiratory symptoms. The most commonly reported symptoms related to compression of adjacent structures by aberrant right subclavian artery are dysphagia , respiratory distress, retrosternal pain (17.0%), cough , and weight loss greater than 10 kg over a 6-month period. Anomaly is clinically silent in 90-93% of cases. Symptoms, when present, occur at the two extremes of life. In children, tracheal obstruction or dysphagia can occur. The increased frequency of pulmonary infections seen in infants is thought to be due to the absence of tracheal rigidity. The most common vascular anomalies coexisting with an aberrant right subclavian artery were truncus bicaroticus, 19.2% ; Kommerell's diverticulum(pouch like aneurysmal dilatation) 14.9% ; aneurysm (just after the origin of arteria lusoria), 12.8%; and right-sided aortic arch, 9.2% .The presence of an aberrant right subclavian artery is also higher in disorders such as Down, DiGeorge, and Edwards' syndromes and Tetralogy of Fallot

Surgical intervention is indicated for all patients who have symptomatic or aneurysmal aberrant RSA. In 1946, Gross9 performed the 1st operation to repair this anomaly. At first, treatment for aberrant RSA consisted of ligation of the vessel. Reimplantation or bypass anastomosis of the divided subclavian artery to the ascending aorta or the right common carotid artery or endovascular occlusion have been tried with success.

Conclusion:

In this baby the aberrant right subclavian artery and the patent ductus arteriosus formed a vascular ring constricting the trachea causing respiratory distress and suprasternal retractions. Our perseverance that with suprasternal retractions alone, it has to be an upper airway obstruction helped to clinch the diagnosis and save the baby. The diagnosis of PPHN was a red herring.

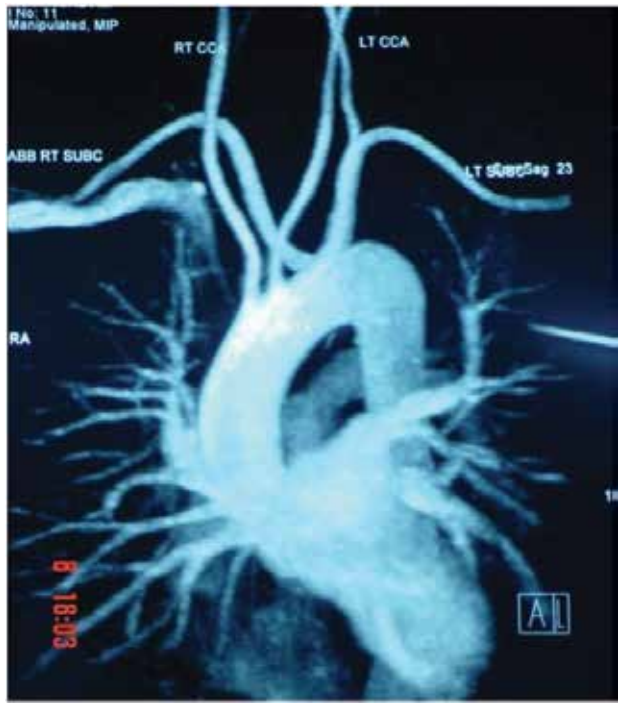

Fig1: Abnormal anomalous right subclavian artery and PDA forming a vascular constriction ring around the trachea

Eventeration of Diaphragm - an unusual response to High frequency Oscillatory ventilation - When to intervene?

Dr Binu Govind, Dr Judy, Dr Arun

Lifeline superspeciality hospital, Adoor, Pathanamthitta

Abstract

Eventeration of diaphragm commonly remains asymptomatic in the newborn period, but some may present with severe respiratory embarrassment. This baby responded well to HFOV as seen in the Xray results, but could not be stabilized for surgery. This brings into debate the need for early intervention in case of severe eventeration of diaphragm.

Introduction

This refers to a case of Eventeration of diaphragm ® side which responded to High frequency oscillatory ventilation (HFOV) but the case deteriorated during the process of stabilization for surgery. Eventeration of diaphragm is an anatomical defect which can be corrected by surgery if the underlying lung is normal. This baby had responded to high Mean Airway Pressure (MAP) in HFOV as the diaphragm was pushed down, however when the pressure was gradually weaned for stabilization, the diaphragm went back to the defective position and could not be taken up for surgery and baby expired.

Case Report

Baby was born at 38 weeks of gestation with a birth weight of 2750g, cried immediately after birth and had secondary apnoea. Cord pH done showed a Ph 7.11, Hco3 17.7 and BE -12 mmol/L. Baby was intubated and brought to nursery and was initially put on conventional ventilation. An X-ray taken showed eventeration of Right Diaphragm [Fig1]. Arterial Blood gas done on Conventional ventilator showed Respiratory acidosis with a Ph 7.12, PCO2 78mmHg However, even with high pressures (PIP28, PEEP5, Fi02 100%), saturation could not be maintained and so mode of ventilation was changed to HFOV with MAP (Mean airway pressure) 28, Δp34 (amplitude) and FiO2 100%. With these parameters, saturation improved and so Fio2 was weaned to 45%. Repeat blood gas done showed a pH7.38, PCO2 38, PO2

76mmHg, HCO3 17mmol/L and BE-8mmol/L. Xray taken showed that the Rt Diaphragm had been pushed down following the high MAP [Fig2]. The underlying lung was well developed. MAP was gently tapered to around a MAP of 22 to prepare the child for surgery, but the baby condition deteriorated and then did not recover. A repeat Xray taken showed that the diaphragm had gone back to its original position [Fig3]. A further increase in pressure did not help in pushing back the diaphragm. Baby condition gradually deteriorated over the next few hours and the baby could not be taken up for surgery. Baby expired at 7 hours of life secondary to hypoxia and associated pulmonary hypertension.

Discussion

Eventeration is a defect usually caused by a deficiency of muscle and it is usually unilateral and commonly on the left side[1,2,3,4]. The frequency of eventeration was found to be 4% and only 3 cases out of them had significant symptoms[5]. In another study, the incidence was reported to be 1 in 1400[6]. The muscles in the peripheral part are well muscularised, whereas the involved part may be sparsely muscularised and covered by an aponeurotic membrane. This is different from diaphragmatic hernia where the defect is complete.

Eventeration may be classified into two- Congenital or Acquired [7,8,9]. Congenital eventeration is of three types- Complete, Partial and Bilateral. Acquired cases may be secondary to injury to phrenic nerve following breech delivery[10]. A significant proportion of these babies will have minimal oxygen requirements or may be asymptomatic[8]. Babies with eventeration may rarely present as acute respiratory distress[10] or difficulty in feeding[11,12].

Some babies may have respiratory distress necessitating intervention Preoperatively, these babies, if they have sufficient respiratory distress and cyanosis, are intubated at sufficient pressures and humidity to maintain oxygenation. The high pressures is supposed to open out the collapsed lung and maintain adequate oxygenation. There are instances where the pressure requirements are high and necessitate the use of High frequency ventilator (HFO). There are studies on preoperative stabilization of diaphragmatic hernia with HFOV as a part of gentle ventilation[13].

In our case, the pressure requirements in HFO were so high that they could not be weaned to a level where surgery could be done within reasonable safe levels. The high levels initially helped in pushing the diaphragm down, but an attempt to wean the pressures resulted in the diaphragm going up and the patient deteriorating.

The appropriate timing of surgery is uncertain. In one study, it is noted that early surgery improves the long term respiratory function[9].

It would be worthwhile considering how much of time it is necessary to wait in situations like these and whether early surgery in babies like these will improve the prognosis, in spite of the increased pressure requirement (MAP). The possibility of the baby deteriorating over the stabilization period has to be considered and the danger of accompanying pulmonary hypertension.

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Immature Anterior Mediastinal Teratoma in a New-born

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Abstract:

Teratomas are neoplasms that originate in Pleuripotent cells, composed of a wide diversity of tissues foreign to the organ or anatomic site which they arise. Here we present a new-born who was delivered with an antenatal diagnosis anterior mediastinal mass which turned out to be an intra-pericardial immature teratoma. This case report explains the difficulty in interpreting the pre-operative diagnostic evaluation in mediastinal lesions, which was finally removed completely at the third thoracotomy leading to full recovery.

Key words: Teratoma, mediastinum, thymus

Case Report:

A full term primigravida mother was referred to our hospital in labour, with antenatal USG showing irregular solid and cystic mediastinal mass (3.1 x 2.1cm). She delivered a 2.3 Kg IUGR female infant by normal vaginal delivery. The liquor was clear. Baby was apneic at birth with APGAR score of 5 & 6 at 1/5 minutes and was resuscitated and intubated from labour room, after which saturation improved and she was shifted to NICU with 40% FiO2. Baby had no dysmorphism or external congenital anomaly and the system examination was unremarkable.

CXR showed large mediastinal mass extending to the right lower chest (fig 1) and CT chest reported as 5.5 x4.16 x2.8 cm lobulated well marginated soft tissue mass in the anterior mediastinum extending from the level of thoracic inlet to the costo-phrenic recess anteriorly. No mediastinal or hilar lymphadenopathy noted and possible diagnosis ofthymoma was suggested. (Fig 2 & 3). Median Sternotomy was done on day5 of life which showed grossly enlarged thymus attached to pericardium and reaching up to midsternal level and extending to the right hemi-thorax. Gland was separated from pericardium and part of it excised for biopsy, which was reported as thymic hyperplasia. Meanwhile S. AFP level came as high (29131ng/ml - normal at day 4 of life is 0.5 -18964) and B HCG was normal. Baby was extubated 2 days later and discharged on oral prednisolone to promote regression of thymic hyperplasia.

2 weeks later baby came back with failure to thrive and cardiac failure. The anterior mediastinal mass had increased in size on repeat CXR and CT scan chest. A repeat surgery with median sternotomy was done on day 51 of life, which showed markedly regressed

thymus with appearance suggestive massive pericardial effusion / multi-loculated pericardial cyst. Pericardium was opened and de-roofed and drained 70ml pericardial fluid. Baby had intra-operative bradycardia and desaturations and hence further exploration could not be done and baby was ventilated post operatively. Post-operative CXR showed reduction in size of mediastinal mass. Biopsy was reported as consistent with pericardial cyst. Pericardial fluid showedmesothelial cells and lymphocytes dispersed in a background of fibrinous material.

Further a 2D Echo showed extra - pericardial multi-cystic mass abutting the right atrium with no apparent communication with pericardial cavity. A repeat CT chest revealed a 5 x 3.5cm multi-loculated cystic anterior mediastinal mass at right paracardiac area. (Fig 4 &5).

A repeat surgery was performed with right thoracotomy, which revealed no extra-pericardial lesion and hence pericardium was opened and 5cm firm mass with cystic areas, attached to the root of aorta was identified, the same was excised completely, there was no infiltration or mediastinal lymph nodes. The feeding vessels were from aorta. (Fig 6). Postoperatively baby was ventilated for 24 hours and had an uneventful recovery. Biopsy was reported as Immature anterior mediastinum teratoma, grade II.

Follow up after 1 year, no recurrence and child is thriving well. S. AFP became normal.

Discussion:

Anterior mediastinal masses in infants and children could originate from thymus (Thymic hyperplasia, thymoma, thymic carcinoma and thymic cyst), lymphoma, lymphatic malformation, teratoma and lipoma). Mediastinal teratomas are rare in infants and children, accounting for 7-11 % of all germ cell neoplasms 1 and intra-pericardial teratomas are even rarer (<1%).[1]

Our baby had both thymic hyperplasia and intra-pericardial teratomawhich is very difficult to differentiate on imaging modalities, unless calcification or other specific structures are identified in the lesion. Surgical excision is the treatment of choice for mediastinal teratomas and a median sternotomy gives best exposure for surgical exploration.[2] Since the first CT report came as thymic hyperplasia, the first surgery was done as median sternotomy and only thymus was explored, pericardium was not opened. Subsequently baby was symptomatic and CT showed pericardial mass and fluid and hence pericardium was opened and explored which showed an intra-pericardial teratoma. Complete removal of the tumour without causing damage to the adjacent structure is often curative in teratoma of infants without recurrence.

Message:

1. Infants with antenatally diagnosed mediastinal mass are best delivered at a tertiary care centre with expertise and facility for immediate respiratory support.
2. The aetiology of anterior mediastinal mass can be often very challenging by imaging modalities.
3. Complete removal of teratoma is mostly curative for infants and recurrence is rare.

Reference:

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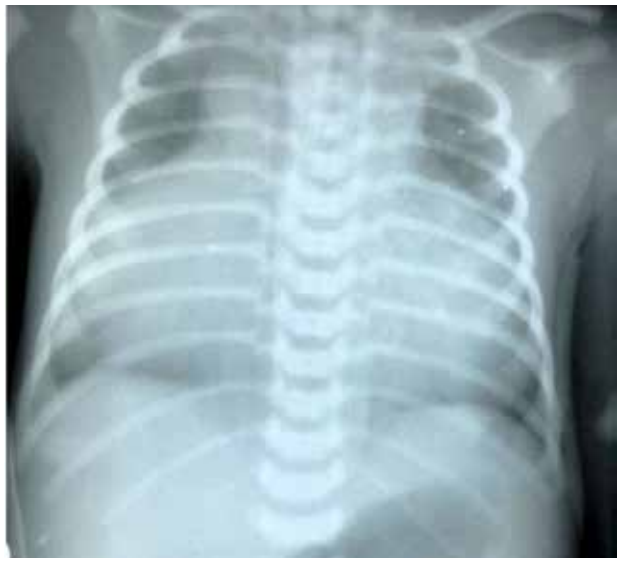

Figure 1: CXR showing large mediastinal mass extending to the right lower thorax.

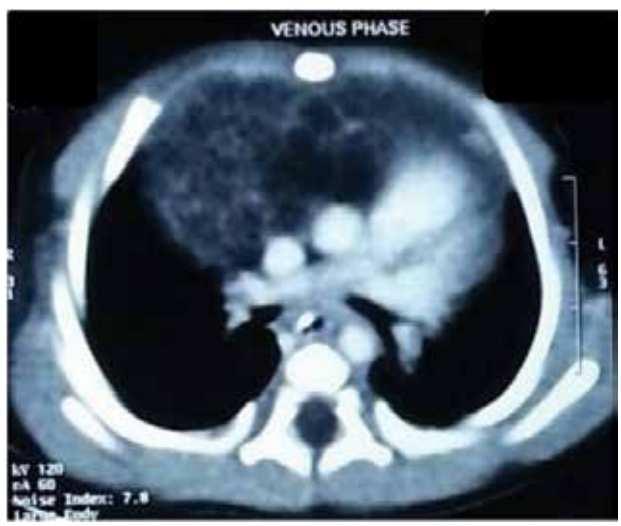

Figure 2. CT scan chest showing the anterior mediastinal mass.

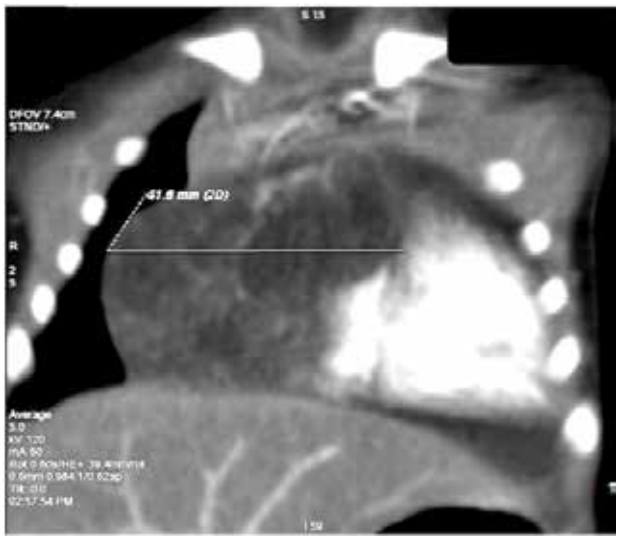

Figure 3. CT scan chest showing the anterior mediastinal mass.

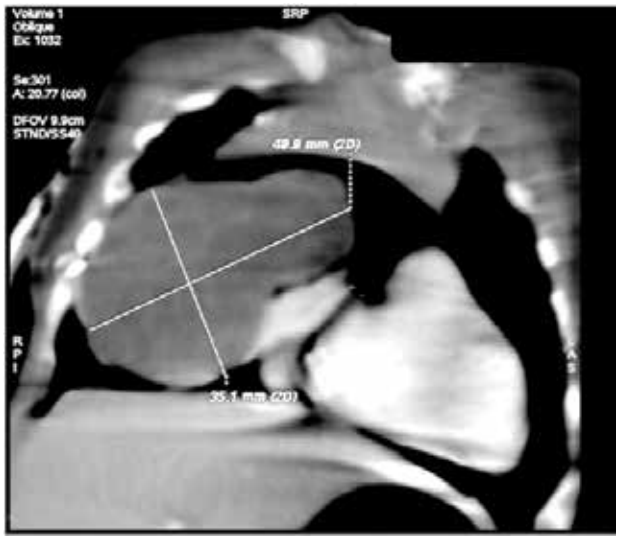

Figure 5. Repeat CT scan showing mass mainly located on the right side of heart.

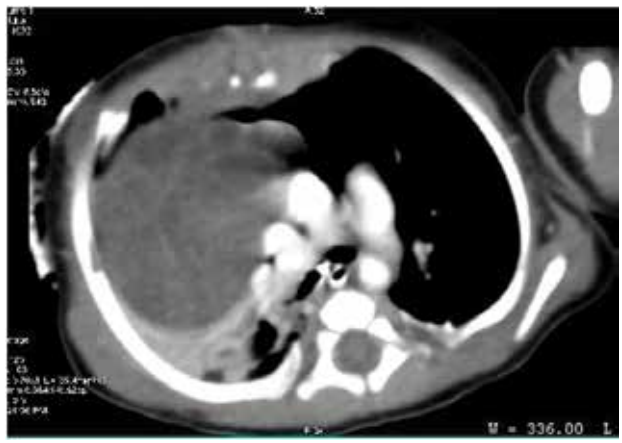

Figure 4. Repeat CT scan showing mass mainly located on the right side of heart.

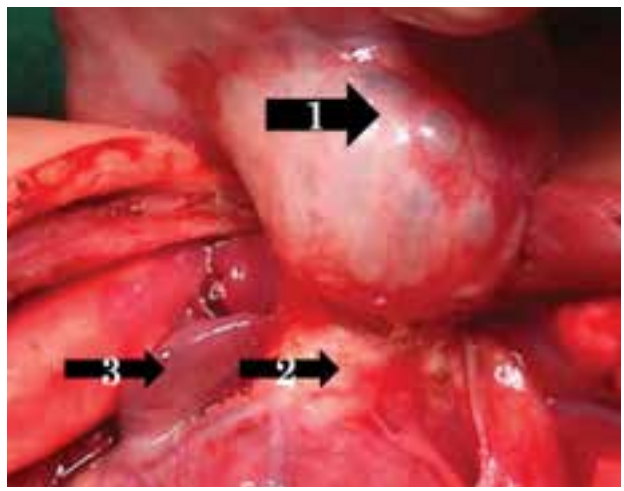

Figure 6. The mass with neighbouring structures identified during surgery.

1. Teratoma
2. Root of Aorta
3. Right Atrium
4. Left ventricle,
5. left Atrium.

Puzzle at Birth

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Case study

A baby weighing 680 grams was born by emergency LSCS, a primigravida who had an uneventful antenatal period. A non - consanguineous marriage and spontaneous conception, 29 -30 weeks gestational age, and SGA.

On examination:-

- Baby's external genitalia examination showed a prominent hyperpigmented phallus with labia. Phallus did not have urethral opening, which was in the perineum.
- This baby was ventilated for respiratory distress for about a week.
- Baby's blood and electrolytes were within normal limits. (Hb- 21 gm%, PCV- 58.9%, TC- 16868 cells/cumm, Platelets - 1.96 Lakh cells, Serum sodium - 144 m Eq/L, S. Pottasium - 5.3 m Eq/L, blood urea - 47 mg/dl, S. creatnine - 1mg/dl, TSH- 1.7 ulU/ml).
- Neurosonogram on day 3 normal.
- Ultrasound abdomen and pelvis showed 2.6mm endometrial shadow. Ultrasound of inguinal canal visualised both gonads in inguinal canal.
- Endocrinology evaluation revealed possible deficiency in 5 alpha reductase
- Subsequent Gonadal biopsy revealed presence of seminiferous tubules.

In the 3rd week, 17 - hydroxy progesterone levels were 21 ng/ml (0.1 - 9.4 ng/ml normal range)

Repeat value of 17 - Hydroxyprogesterone (at 1 month) was 3.73 nmol/L

Chromosomal analysis - Male Karyotype

At 1.5 years of age

	Bio. Ref. Level
(5alphadihydrotestosterone observed value 54.7 Pq/ml	< 30
Androstenedione (below 0.3 ng/ml	0.05 - 0.45
Testosterone <12.98 ng/dl	Prepubertal < 30

Ambiguous genitalia in the Newborn

- Ambiguous genitalia is a significant example of a disorder of sexual development in which the external genitalia do not have the typical appearance of either sex.
- Diagnosing a newborn with DSD or ambiguous genitalia should be treated with urgency and the Neonatologist on call should be informed.
- One of the first questions asked of and by parents is whether their baby is a boy or a girl. It is understandable that most families prior to diagnosis of DSD will not even have considered that gender could be ambiguous. Therefore not only do they have the loss their ' Normal' child to grieve for, they also have extra stress of what to say to family and friends.

What to do ?

1. Be empathetic & sensitive
2. Don't make any comments that could be misinterpreted as indicating a gender
3. Keep initial discussions short and simple
4. Examine the child's genitals in the parents presence and use gender neutral language.
5. Discuss the ongoing plan and follow up; ensure a multidisciplinary discussion with Endocrinology and genetics and pediatric surgery

Relevant questions to ask family

1. Drug ingestion, infection or exposure to teratogens during pregnancy
2. Any recent androgenic changes in mother suggesting androgen excess
3. History of consanguinity (suggesting autosomal recessive inheritance)
4. Previous siblings dying in newborn period
5. Previous siblings with over virilisation or precocious puberty

Useful examination findings

1. Signs of hypoglycemia or dehydration
2. Palpable gonads in the labioscrotal or inguinal regions
3. Penile length and width normal (Normal 2.5 - 4.5cm in full term infant)
4. Position of urethral opening
5. Labioscrotal fold fusion
6. Genitalia pigmentation
7. Syndromic features or other physical abnormalities

Investigations

1. Serum electrolytes and Glucose
2. Chromosome analysis (this needs to be asked for urgently - a FISH for chromosome can be provided within 48 hours).
3. Pelvic/abdominal ultrasound to determine absence or presence of a uterus. This requires an experienced sonographer (A uterus almost always indicates no functioning testes are present while no uterus indicates functioning sertoli cells and therefore testicular tissue)
4. 17 - hydroxyprogestone (Newborn screening card for urgent 17OHP)

Gender assignment

Gender identity development is the result of a complex interaction between genes and environment. It is difficult to predict what gender the child will eventually identify with.

The role of health care professionals in initial gender assignment is

1. To obtain and interpret test results of
   a. The etiology and prognosis of the child's DSD
   b. Concerning the status of the child's anatomy and physiology,
2. To inform the parents and assist them in the decision about gender assignment.

Collodion Baby

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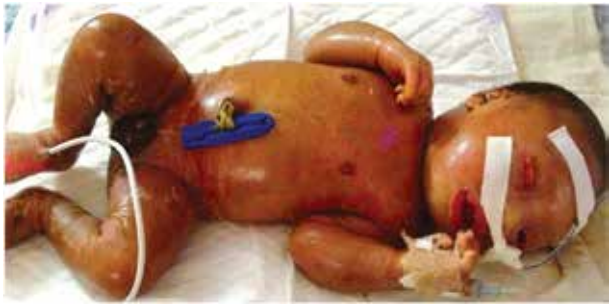

(Dr. Johny VF, Kochi)

(Dr. Johny VF, Kochi) Marked clinical improvement after 2 weeks

Essential Features:

- It is a manifestation of congenital ichthyosiform erythroderma or lamellar ichthyosis.
- They look like oiled parchment or collodion at birth due to the thick and taut skin.
- They have, flattening of the ears and nose and fixation of the lips in a O - shaped configuration.
- Sometimes the infant may develop normal skin after the shedding of this thick membrane.
- Treatment is mainly non-occlusive lubricants and a high humidity environment.

Description:

The collodion baby is a descriptive term for the infant who is born encased in a tight shiny membrane that resembles plastic wrap. The collodion baby is not a disease entity but is the first expression of some forms of ichthyosis.

Clinical picture:

The collodion membrane cracks and peels over the course of several weeks. The tightness of the membrane may cause an ectropion. Eclabium, the turning out of the lips due to the tightness of the membrane, may accompany the ectropion, and may cause difficulties with nursing. When the membrane is completely shed the infant may display one of several ichthyosis skin types. Congenital ichthyosiform erythroderma (CIE) and lamellar ichthyosis are the most commonly seen forms of ichthyosis presenting with a collodion membrane. A small percentage of infants shed the membrane and never display any other skin involvement; a phenomenon called “self-healing collodion baby.”

Complications:

The cracking and peeling of the membrane increases the risk of infection. These infants are also at risk for fluid loss, dehydration, electrolyte imbalance, body temperature instability, and pneumonia.

Management:

Collodion babies should be placed in a high humidity chamber, and monitored closely for complications. A high humidity environment will allow slow, gradual sloughing off of the membrane. The membrane will come off on its own and should not be peeled off. Application of mild petroleum-based moisturizers may help the infant feel more comfortable while the membrane is peeling off.

Hallermann - Streiff Syndrome

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Essential Features:

- Hallermann-Streiff Syndrome is a rare genetic condition which presents with congenital anomalies of head and face.
- Also called dyscephalia mandibulo-oculo-facial syndrome.
- The characteristic features include: dyscephalia and bird face, dental anomalies, proportionate short stature, hypotrichosis, atrophy of the skin, bilateral microphthalmia and congenital cataracts.
- They are mostly sporadic and usually not associated with chromosomal anomalies.

The outstanding features of the syndrome are as follows:

- Dyscephaly with bird-like face and hypoplastic mandible
- Proportionate short stature
- Congenital cataracts
- Localized hypotrichosis
- Congenital abnormalities of the eyes
- Cutaneous atrophy limited to face
- Mental retardation

Aetio-pathogenesis:

The aetiology of this rare genetic condition is attributed to an asymmetric second arch defect that develops between the fifth or sixth gestational week. Almost all cases are sporadic and only few cases have demonstrable chromosomal anomalies. There are some recent reports of defects in elastin and glycoprotein metabolism.

Investigations:

Thorough ophthalmologic, ENT and dental assessment should be done in all patients

Complications:

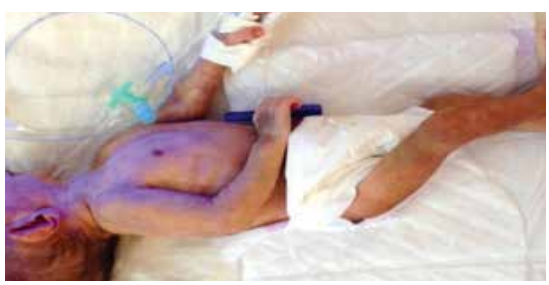
Upper airway obstruction due to small nares and glossoptosis secondary to micrognathia, may lead to cor pulmonale. Tracheomalacia can lead to chronic respiratory insufficiency, resulting in biventricular cardiac failure and early death. Developmental failure of the skeleton, delayed bone age, failure of the rib and clavicle, spina bifida and scoliosis can occur.

Management:

There is no cure for this syndrome. Affected individuals need periodic ophthalmologic, ENT and dental assessment and interventions. Genetic counselling must be offered to all affected individuals and their families.

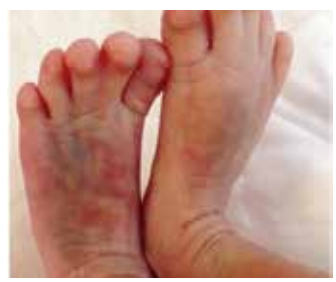
Prognosis:

Individuals affected with Hallermann-Streiff syndrome may have normal intelligence and life span when complications of this disorder are properly managed. Early death due to respiratory difficulties is known to occur.



(Dr. Johny VF, Kochi)

(Dr. Johny VF, Kochi)
Congenital cataract



(Dr. Johny VF, Kochi)
Syndactyly of 2nd and 3rd toes bilaterally

Rubinstein Taybi Syndrome

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Essential Features:

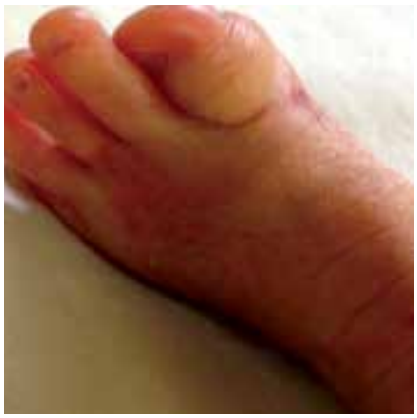
- Infants with Rubinstein-Taybi syndrome have thumbs and/or great toes that are abnormally broad as a result of unusual broadness of the terminal phalanges. In addition, distal bones of the thumbs and great toes may also be angled improperly (misaligned) on a proximal bone that is abnormally shaped (delta phalanx). The fifth fingers may be permanently fixed in a bent position (clinodactyly).
- Most affected infants experience varying degrees of mental retardation, delay in the acquisition of skills requiring coordination of muscular and mental activities (psychomotor retardation), and delayed socialization.
- Infants with Rubinstein-Taybi syndrome may have a wide variety of distinctive craniofacial abnormalities, like an abnormally large, "beak-shaped" or straight nose with a broad nasal bridge, a peculiar grimacing facial appearance when smiling and laterally downslanting palpebral fissures, as well microcephaly below the 50th percentile, with an unusually prominent forehead (frontal bossing).

(Dr. Johny VF, Kochi)

(Dr. Johny VF, Kochi)
Rubinstein Taybi Syndrome-Distinctive facial features.

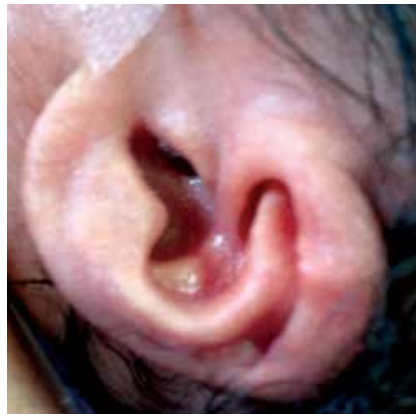
(Dr. Johny VF, Kochi)

Rubinstein Taybi Syndrome-Broad thumb.



(Dr. Johny VF, Kochi)

Rubinstein Taybi Syndrome-Broad great toe.



(Dr. Johny VF, Kochi)

Rubinstein Taybi Syndrome-Dysplastic pinna.

Rubinstein-Taybi syndrome

Rubinstein-Taybi syndrome is a rare genetic multisystem disorder that typically affects many organ system of the body. The group of physical findings and symptoms associated with this syndrome include distinctive abnormalities of the fingers and toes, developmental delays, growth retardation, speech delays, mental retardation, craniofacial dysmorphism, breathing and swallowing difficulties, skeletal malformations, and/or urogenital abnormalities. In many cases, the skin, heart, and/or respiratory system may also be affected.

Aetio-pathogenesis:

In most cases, Rubinstein-Taybi syndrome occurs randomly, with no apparent cause. In some cases, a positive family history has been identified that has suggested possible autosomal dominant inheritance. The range and severity of symptoms may vary greatly among affected family members (kindreds). It affects males and females in equal numbers.

Investigations:

Rubinstein-Taybi syndrome is usually a clinical diagnosis, based on characteristic physical findings (e.g., low percentile for length, weight, and head circumference, characteristic facial features, etc.). The diagnosis may be further confirmed by x-ray studies that may reveal characteristic malformations of the bones of the hands and feet.

In approximately 15-20 percent of cases, cytogenetic and molecular study of the CREB binding protein gene region of chromosome 16 can help confirm diagnosis. Because of the possibility of associated congenital heart defects, a thorough cardiac evaluation may be beneficial.

Management:

The treatment of Rubinstein-Taybi syndrome is directed toward the specific symptoms that are apparent in each individual. Affected individuals may require early intervention to prevent and/or monitor respiratory and feeding difficulties. Orthopedic techniques, orthopedic surgery, physical therapy, and/or other supportive techniques may help treat certain associated skeletal abnormalities. Language/speech therapy and augmentative and/or alternative communication techniques are recommended.

Early intervention is important to ensure that children with Rubinstein-Taybi syndrome reach their potential. Special services that may be beneficial to affected children may include special remedial education, special social support, and other medical, social, and/or vocational services.

Families may benefit from contacting parent support groups. Genetic counseling will also be of benefit for affected individuals and their families. Other treatment for this disorder is symptomatic and supportive.

Sirenomelia

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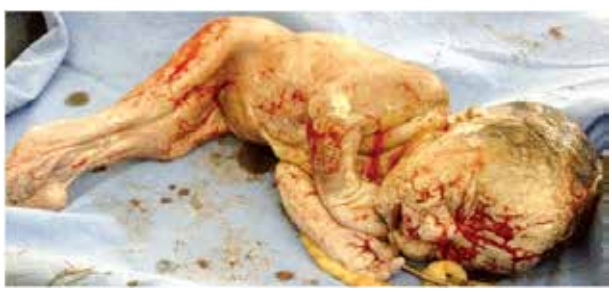

(Dr. Johny VF, Kochi)

Essential Features:

- Sirenomelia is a rare form of caudal dysgenesis, which is generally incompatible with life due to severe kidney malformations associated with it.
- It is characterized by single or fused lower limbs associated with other severe anomalies like bilateral renal agenesis
- Sirenomelia has been classified into three types
  a) Simpus Apus: No feet, one tibia, one femur
  b) Simpus Unipus: One foot, two femur, two tibia, two fibula
  c) Simpus Dipus: Two feet and two fused legs (flipper like)-this is called a mermaid

Aetio-pathogenesis:

Although the precise etiology of sirenomelia is not understood, it is thought to occur due to an embryonic injury between 28-32 days of life involving the caudal mesoderm. Maternal diabetes is a significant risk factor, wherein sirenomelia can occur as a part of the caudal regression syndrome. Exposure to teratogens, such as retinoic acid, cadmium, cyclophosphamide, cocaine and the antiepileptic drug lamotrigine, are also associated with risk of developing sirenomelia.

Investigations:

Prenatal diagnosis of sirenomelia is possible by high resolution or color Doppler sonography, which demonstrate oligohydramnios, bilateral renal agenesis, a single lower limb, a unique umbilical artery, absence of a bladder, undetermined external genitalia, anorectal atresia and lumbosacral agenesis. One of the most important early findings in prenatal imaging is a single umbilical artery of abnormal origin, with single umbilical artery of vitelline origin being considered characteristic of sirenomelia. Postnatally, the diagnosis is confirmed by the distinctive physical features and radiographic findings.

Management:

Medical termination of pregnancy should be offered to all antenatally diagnosed cases, as it is nearly universally fatal after birth. Managing sirenomelia is difficult and quite costly, requiring several surgical interventions for the associated genitourinary and gastrointestinal anomalies. Post-natal management requires the presence of kidneys, even if they are dysgenesic.

Prognosis:

Sirenomelia is fatal in most cases because of the characteristic pulmonary hypoplasia and renal agenesia. There are exceptional reported cases of survivors, who require extensive surgical interventions.

Protecting The Premature Brain… Current evidence based strategies

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Improving neurodevelopmental outcome for preterm infants is an important challenge for neonatal medicine. It could be done at various levels:

- Antenatal interventions
- Postnatal Interventions
- Preventing causative morbidities
- Emerging strategies

ANTENATAL INTERVENTIONS

- In- utero transport of anticipated preterm delivery
- Antenatal steroids
- Antenatal magnesium sulphate
- Management of PPROM

ANTENATAL TRANSFER FOR ANTICIPATED PRETERM DELIVERY

If a preterm delivery is anticipated, then the best transport mode is in utero. Always try to deliver a preterm baby in a hospital where there is level 3 neonatal facility, when feasible.

Transport during first 48 hours carries risk of IVH. Tertiary centre care has shown better morbidity free survival. Hence "care centralisation" is important. A co-ordinated neonatal & obstetric network strategiesis needed for safe antenatal centralisation.

ANTENATAL STEROIDS

Antenatal steroids have been recommended for all women with threatened preterm delivery before 34 weeks gestation. It has shown to decrease the incidence of IVH & white matter damage. Lower rates of developmental delay and cerebral palsy and higher cognitive ability have been noted in preterm infants whose mothers received antenatal steroids

Current recommendation for single versus multiple courses of antenatal steroids are not clear. Repeat courses may be used if delivery has not occurred within 7days. Multiple steroid courses[1] has the advantage of lower risk of early respiratory morbidity. However disadvantages include reduced birth weight & head circumference. Current guidelines[3] advise single complete course. Single additional rescue course may be appropriate when the first was before 26 weeks gestation.

Betamethasone is currently recommended by RCOG and ACOG. At present, there is no significant evidence to support superiority for either of them. There have been 10 Cochrane reviews so far regarding betamethasone versus dexamethasone.

MAGNESIUM SULPHATE

It is a potential neuroprotectant. Studies show reduced incidence of IVH and better neurodevelopmental outcome. Current recommendations suggest $MgSO_4$ for all deliveries prior delivery prior to 32 weeks gestation.

Dose is 4 g loading dose over 20 - 30 min followed by an infusion of 1g / hour until birth or for a maximum of 24 hours.

MANAGEMENT OF PPROM

It is important to manage PPROM. Aim should be to reduce chorioamnionitis & early onset sepsis which has shown to

cause adverse neurodevelopmental outcome due to cerebral hypoperfusion, capillary thrombosis, increased permeability of blood brain barrier, direct passage of microbial products & proinflammatory cytokines into cerebral tissue

However, there are controversies regarding prophylactic antibiotics versus immediate delivery after 34 weeks

Prophylactic antibiotics have shown to prolongation of pregnancy, reduction in neonatal infection and fewer abnormal NSGs

Recent RCTs[7] have shown no difference in incidence of sepsis but increase in preterm complications making recommendations for immediate delivery contentious

POSTNATAL INTERVENTIONS

- Deferred cord clamping
- Caffeine for apnoea of prematurity
- Indomethacin prophylaxis for PDA
- Volume targeted ventilation to prevent hypocarbia

DEFERRED CORD CLAMPING

Studies have shown reduction in mortality & morbidity (IVH) following deferred cord clamping. It has also shown to have improved-motor function at 18 -22 months[8]. The effects may be due to increased blood volume &oxygenation, prevention of anaemia and transfer of stem & progenitor cells with extensive proliferative capacity- thereby repairing tissues & promoting immunocompetence.

But there is always a hesitancy because of lack of consensus on optimal timing, risk of volume overload, polycythemia

A recent study by McAdams et al, has shown that delayed cord clamping is feasible, safe and have significant benefit to preterm with no detriment from the risks above.

CAFFEINE

Recurrent apnoeashave been shown to be harmful to a preterm baby. CAP trial has shown decreased incidence of cerebral palsy & cognitive impairment at 18 - 21 months when treated with caffeine in first 10 days. It is safer than other methyl xanthines and is recommended.[9]

Caffeine when given within 72 hours has shown reduction in bronchopulmonarydysplasia, PDA and mortality

INDOMETHACIN PROPHYLAXIS FOR PDA

Indomethacin prophylaxis has shown to reduce the incidence of severe IVH (grade 3 & 4), ventriculomegaly and periventricular leukomalacia. This may be due to the direct effect on brain by reduced prostaglandin synthesis, reduced cerebral vascular hyperaemic response and improved maturation of basement membrane & basal lamina. There is also reduced surgical need for PDA. Babies without PDA are less vulnerable to hypoxic, hypercapneic, hypertensive insults.

VOLUME TARGETED VENTILATION TO PREVENT HYPOCARBIA

Using newer modes of ventilation has shown to improved neurodevelopmental outcome in preterm babies. This could be due to prevention of hypocarbia andchanges in cerebral blood flow & perfusion pressure.

A recent meta-analysis volume targeted Vs pressure limited ventilation revealed significant reduction in hypocarbia and thus reduction in PVL & grade 3-4 IVH

PREVENTING CAUSATIVE MORBIDITIES

- Late onset sepsis
- Necrotising Enterocolitis
- Poor nutritional status

LATE ONSET SEPSIS

In any neonatal unit, strict aseptic precautions are necessary . Standardised care bundles for central lines, Judicious use of antimicrobials, limited postnatal steroid use, early enteral feeding has all shown to reduce late onset sepsis.

Studies are ongoing to assess the usage of bovine lactoferrin supplementation, probiotics and immune replacement therapy

NECROTISING ENTEROCOLITIS

Promotion of breast milk and avoidance of bovine origin products has been proven beyond doubt in reducing NEC. PIPS trial has suggested probiotic usage in preterm infants may reduce NEC.

EMERGING STRATEGIES

Newer neuroprotective strategies under research include use of melatonin, erythropoietin and stem cell therapies.

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The Face that Predicts the Brain

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Holoprosencephaly (HPE)

Holoprosencephaly (HPE) is the most frequent malformation of the prosencephalon. It represents the absence or incomplete division of the prosencephalon during the 4th and 8th week of gestation. Its incidence is estimated to be 1 in 16000 live births and 1 in 250 spontaneous abortions. It is classified in 3 types, according to the degree of cerebral involvement: alobar, semilobar and lobar. The clinical features vary very much, depending on the severity of holoprosencephaly.

We present a 3 week old neonate diagnosed with holoprosencephaly and give a brief discussion on the pathogenesis, clinical features, management and prognosis of holoprosencephaly.

CASE REPORT

A 3 weeks old male infant, first born of non consanguineous parentage.

Born at term out of a NVD, cried soon after birth but developed respiratory distress.

The maternal history was unremarkable for any comorbid conditions; prenatal infections or any other chronic disease. She was not on any medications.

No significant family history was elicited.

Evaluation revealed bilateral Choanal Atresia , was operated on day 3 and was under periodic nasal dilatation.

Baby now presented with incessant cry and opisthotonic posturing since 4 days.

CT brain suggested partial corpus callosal agenesis

Baby was referred for further evaluation

O/E-: pale, emaciated baby in opisthotonic posturing Microcephaly, dysmorphic with prominent eyes, long upturned eyelashes & midfacial hypoplasia

Hypertonic with clenched fist

Anthropometry - HC -32.5 cm, Weight - 2.9 kg

Systems:

Respiratory system	:	Audible laryngeal stridor
Cardiovascular system	:	S1S2 heard, normal pulses
Per abdomen	:	soft, non tender, no organomegaly
Central nervous system	:	AF-normal, Increased tone in all limbs, dystonic posturing

Evaluation :

MRI brain -

Hypoplasia of bilateral frontal lobes with partial fusion and abnormal gyral pattern, partially fused thalami, slit like 3rd ventricle, absent frontal horns and partial agenesis of corpus callosum suggesting SEMILOBAR HOLOPROSENCEPHALY

Further Workup:

TFT - normal
Serum cortisol- normal
ILGFBP -3 - borderline low
Serum lactate - mildly elevated
Lactate: pyruvate ratio - elevated
EEG - normal
BERA – normal

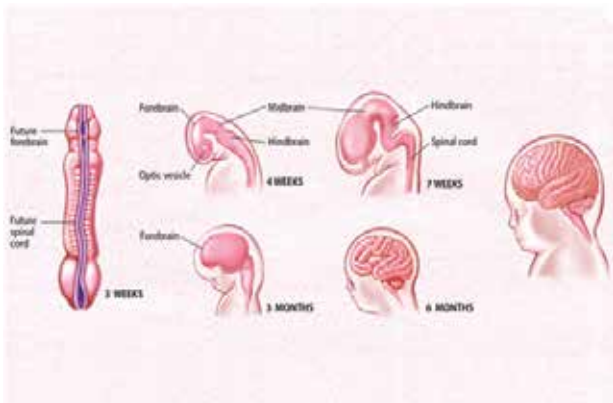
HOLOPROSENCEPHALY

Holoprosencephaly is a complex intracranial abnormality characterized by absent or incomplete cleavage of prosencephalon

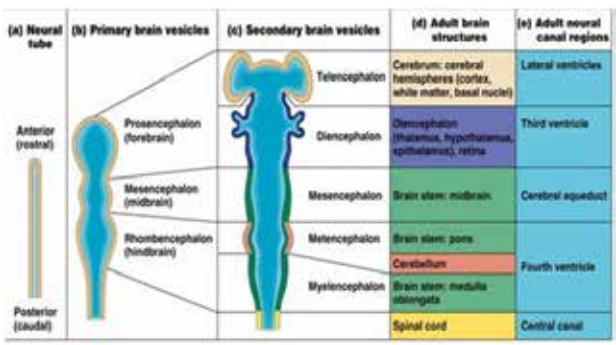
Key facts :-

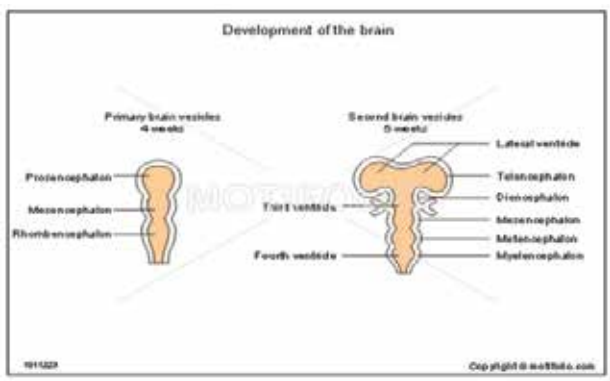
- A disorder of gastrulation
- Most common structural anomaly of developing forebrain
- Occur at 2 – 3 weeks post conception
- Incomplete midline clevage of prosencephalon
- Neurologic impairment and dysmorphism of brain and face
- Observed in 1 :250 conceptuses
- High rate of fetal demise
- Birth prevalence is 1:8000 live birth
- Heterogenous eitiology

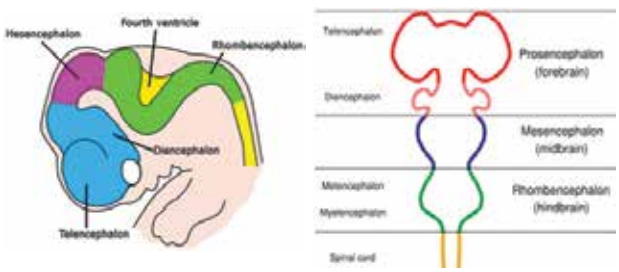
Note – CNS development – occur in third week of life
Gastrulation – cell movement
spherical ball of cells - mulitilayered
Neurulation – nervous system from ectoderm



ANATOMY OF THE DEVELOPING BRAIN







Holoprosencephaly (HPE) represents an incomplete or absent division of the prosencephalon (forebrain) into distinct cerebral hemisphere usually occurring between 18th and 28th day of gestation

TYPES OF HOLOPROSENCEPHALY:

Divided into four

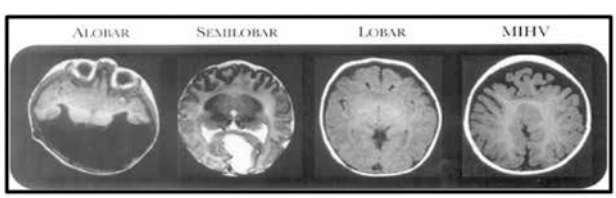
- A disorder of gastrulation
- Most common structural anomaly of developing forebrain
- Oc
- Based on degree of non separation of prosencephalon

  Alobar form - Diffuse cortical

  Semilobar form - Frontal lobes

  Lobar form - Basal aspect of frontal lobes

  Middle interhemispheric variant - Post frontal and parietal lobes

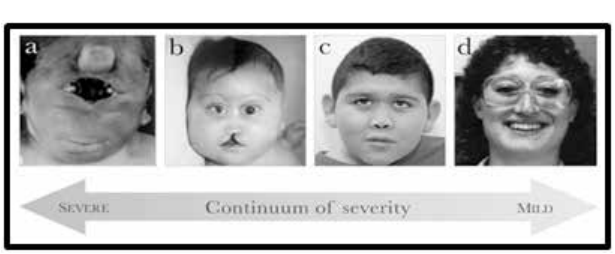


- Severity and prognosis - Depend on degree of non separation

  Alobar - most severe, microforms

  Severe form - Pronounced microcephaly

  Cyclopia, synopthalmia. proboscis



ETIOLOGY

- Environmental factors and teratogens:

  Genetic causes, maternal DM , maternal hypocholestrolemia, Ethanol ,CMV infection
- Syndromic association:

  Smith lemili opitz syndrome, Pallister Hall syndrome, Rubinstein taybi syndrome, Meckel Gruber sundrome
- Chromosomal anomalies: 24 - 45 % of live births affected by holoprosencephaly

  Most frequent numeric anomalies in chromosome 13,18,21

  Structural anomaliesnvolving 13p, 18p, 7p36, 3p24 -pter, 2p21, 21q 22.3

  Iatrogenic mutation in 4 genes -SHH (7q36), SIX3(2p21), ZIC2(13q32), TGIF (18P11.3)

Note:

Recurrence risk after an isolated case of holoprosencephaly with normal chromosome is 5.6%

Diagnosis - Clinical diagnosis
Neuroimaging
Syndrome evaluation
Cytogenetics
Genetic counselling

Prognosis - Frequent cause of death -respiratory infection
Dehydration secondary to uncontrolled DI
Intractable seizures
Sequalae of brainstem malfunction

Clinical management:

The treatment of HPE is supportive and is oriented towards different malformations associated.

Impaired homeostatic function - Temp. thirst, appetite, sleep wake cycle

Pituitary dysfunction - Posterior - Central DI
Anterior - Hypothyroidism, Hypocortisolism

Motor impairment - Hypotonia, dystonian spasticity, Baclofen and trihexyphenydyl

Oromotor dysfunction -

Cleft lip and palate - aspiration n respiratory infection

Respiratory issue - chronic liver dysfunction

Gastointestinal issues - poor gastric and colonic motility and GER gastrotomy tube, medication and antireflux procedure

Seizures - Complex partial

Hydrocephalus - VP shunt

Conclusion

Holoprosencephaly is the most common forebrain developmental anomalies. The cause can be heterogenous, including a teratogenic and or a genetic basis. It is important to diagnose holoprosencephaly prenatally and determine the type to classify severity, complications and survival rate. MRI is the best modality for diagnosing and classifying the type of HPE.

The babies diagnosed with holoprosencephaly need multidisciplinary treatment approach.

The parents should be counseled regarding the poor prognosis and should be referred to early intervention for physical and occupational therapies.

If you have to have an inherited metabolic disease, then this is the one to have ! : A case report

Dr. Khais K, Dr. Divianath, Dr. Vishnu Mohan, Dr. Anand MR, Dr. Preetha Remesh

Three weeks old male infant with increased jitteriness noted from one week of age & myoclonic jerks noted from 2 weeks of age, that is refractory to AED .Mum reports poor activity from day one 'Baby always sleeping'…But baby sucking well at breast & with good weight gain 3rd child of a non consanguineous marriage; Uneventful antenatal & natal history Born at term weighing 3.7kg & cried soon after birth.

- Elder sibling died at 2 months with a sudden onset breathlessness ? Cardiomyopathy
- On examination
  - Obtunded
  - Dry scaly skin
  - Excessive startle reflex
  - Hyper reflexia

Possibilities entertained at this stage:-

- Malformations of Cortical development
  - Lissencephaly; Focal cortical dysplasia
- Early Myoclonic Encephalopathy
  - Ohtahara syndrome; West syndrome
- Mitochondrial Encephalopathy
- Inherited disorders of Metabolism

Baseline Investigations:-

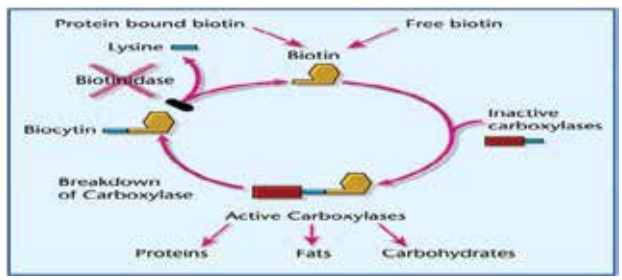
- CRP<5 mg/L
- Electrolytes :- Normal
- Blood gases:- normal
- Lactate : Pyruvate ratio :- <20(normal)
- EEG :- Burst suppression pattern
- MRI brain :- normal
- IEM Work up:- Normal but for elevated 3 OH Iso-valeryl-carnitine C5OH :-6.24 (0.05-1.0)

We are therefore looking at a metabolic encephalopathy without metabolic Acidosis and with an elevated level of 3 OH Iso-valeryl-carnitine

- Confirmatory Test:- Biotinidase level :-7.34 u (normal is > 40)

Biotinidase Deficiency:-

- Autosomal recessive disorder with an incidence of 1/60,000.
- BTD gene is on Chromosome 3p25
- Biotin is a cofactor for carboxylation reactions in gluconeogenesis,Fatty acid metabolism & Amini acid metabolism. Biotinidase is essential for recycling of biotin from protein.



Clinical Presentation:-

- Presents as early as 1 week & up to 10 yrs
- Neurological:-
  - Hypotonia and seizures
  - Developmental delay
  - Respiratory :-hyperventilation, apnoea
  - Sensory neural hearing loss
  - Dry scaly eczematous skin
- Biochemical:-
  - Metabolic Acidosis with ketosis
  - Hyperammonemia
  - Organic aciduria

BUT, as in our baby, all the biochemical changes are relative &absence of these should never rule out the possibility of biotinidase deficiency

Treatment :- free biotin (5-20 mg/24 hr)

- Biochemical changes and seizures quickly respond
- Hearing loss can be averted but if already present, may be irreversible

Our baby was started on Biotin; sensorium improved and seizures are now controlled.

Close neuro developemental monitoring mandatory here. Baby has bilateral SNHL already

"if you have to have an inherited metabolic disease biotinidase deficiency is the one to have"

Genetics in Medicine (2012)14,565-575 doi: 10.1038 /gim. 2011. 6

Postscript:-

Our Baby is now 6 months old, seizure free, on just Biotin. SNHL persisting.

Clinical Clues to Biotinidase deficiency:- CNS symptoms + dry scaly skin + SNHL


PEDIATRICS
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Divisional round of NNF Nursing Quiz 8/8/2017
Hosted by Calicut NNF

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