

**STANDARD TREATMENT
GUIDELINES
PEDIATRICS & PEDIATRIC
SURGERY**

**Ministry of Health & Family Welfare
Govt. of India**

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PROTOCOL FOR DENGUE FEVER IN CHILDREN

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Dengue Fever

1 Introduction

Dengue has a wide spectrum of clinical presentations, often with unpredictable clinical evolution and outcome. Reported case fatality rates are approximately 1%, but in India, Indonesia and Myanmar, focal outbreaks away from the urban areas have reported case-fatality rates of 3-5%.

2 Classification and Case definition

The Newer WHO Classification of Dengue is practical from the management perspective and involves 2 categories- Dengue and Severe Dengue [including both the previously classified categories Dengue Shock Syndrome and Dengue Haemorrhagic fever]¹

Case definition of Dengue fever (DF) ^(1,2):

Dengue fever is an acute febrile illness with one or more of the following:- Headache, retrorbital pain, myalgia, arthralgia, rash, hemorrhagic manifestations, and leukopenia and lab confirmation by ELISA.

Case Definition of Severe Dengue

Severe dengue should be considered if the patient is from an area of dengue risk presenting with fever of 267 days plus any of the following features:

ÉThere is evidence of **plasma leakage**, such as:

- ó high or progressively rising haematocrit;
 - ó pleural effusions or ascites;
 - ó circulatory compromise or shock (tachycardia, cold and clammy extremities, capillary refill time greater than three seconds, weak or undetectable pulse, narrow pulse pressure or, in late shock, unrecordable blood pressure).
- ÉThere is **significant bleeding**.
- ÉThere is an **altered level of consciousness** (lethargy or restlessness, coma, convulsions).
- ÉThere is **severe gastrointestinal** involvement (persistent vomiting, increasing or intense abdominal pain, jaundice).
- ÉThere is **severe organ impairment** (acute liver failure, acute renal failure, encephalopathy or encephalitis, ARDS or other unusual manifestations.)

Warning Signs in Dengue Fever *

- ÉAbdominal pain or tenderness
 - ÉPersistent vomiting
 - ÉClinical fluid accumulation
 - ÉMucosal bleed
 - ÉLethargy, restlessness
 - ÉLiver enlargement >2 cm
 - ÉLaboratory: increase in HCT concurrent with rapid decrease in platelet count
- *(requiring strict observation and medical intervention)

3 Differential diagnosis

Conditions that mimic the febrile phase of dengue infection

- Influenza,
- Measles,
- Chikungunya,
- Infectious
- Mononucleosis ,
- HIV Seroconversion Illness
- Rubella,
- Scarlet Fever,
- Meningococcal infection,
- Drug reaction
- Enteric infections
- Meningo/encephalitis
- Febrile seizures

Conditions that mimic the critical phase of dengue infection

- Acute gastroenteritis,

- Malaria,
- Leptospirosis,
- Typhoid,
- Typhus,
- Viral hepatitis,
- Acute HIV, seroconversion illness,
- Bacterial sepsis, septic shock
- Malignancies Acute leukaemia and other malignancies

4 Investigations

Situation 1

Diagnostic Methods

Test	Advantage	Disadvantage
Antigen Detection [NS-1]	<ul style="list-style-type: none"> • Easy to perform • Early detection 	<ul style="list-style-type: none"> • Not as sensitive as above
IgM assay	<ul style="list-style-type: none"> • Less expensive • Easy to perform • Useful in outbreaks 	<ul style="list-style-type: none"> • May miss secondary infection due to undetectable IgM • Useful only after 5 days • Cross reactivity

Supportive Tests

- Complete Blood Count-Hemoglobin, haematocrit, White Cell Count, Platelet count.
- Blood grouping and Cross Matching.
- Peripheral smear: Indicates the type of anemia and confirms leukopenia
The presence of giant platelets and clumps is indicative of good platelet function
- Serum electrolytes, Urea, Creatinine, Random Blood Sugar
- Liver and renal function tests.
- Chest X Ray

5 Treatment

Situation 1

Out Patient Management

Simple Dengue Fever with no warning signs, adequate oral intake and clinically well.

Home Care Advice

É Adequate rest

É Adequate fluid intake - Milk, fruit juice, electrolyte solution (ORS) and barley/rice water.

É Paracetamol [Acetylsalicylic acid, Mefenemic acid, ibuprofen or other non-steroidal anti-inflammatory agents (NSAIDs) and steroids to be avoided.]

É Tepid sponging

É To look for mosquito breeding places in and around the home and eliminate them

É Antibiotics are not necessary.

To observe for the following **Danger signs** and report immediately for hospital admission

- Bleeding:
 - red spots or patches on the skin
 - bleeding from nose or gums
 - vomiting blood
 - black-coloured stools
 - heavy menstruation/vaginal bleeding
- Frequent vomiting
- Severe abdominal pain
- Drowsiness, mental confusion or seizures
- Pale, cold or clammy hands and feet
- Difficulty in breathing

Out -patient laboratory monitoring- as indicated

- Haematocrit
- White cell count
- Platelet count

Admission Criteria to Secondary Centre

- Child having high fever, poor oral intake, or any danger signs as enumerated above.
- If platelet count < 100,000 /cu.mm or rapidly decreasing trend.
- If haematocrit is rising trend.

- Special Social Circumstances (living far from a health facility without reliable means of transport).

Admission Criteria to Tertiary Centre directly from OPD

- If signs of severe dengue or warning signs.
- If $< 50,000/\text{cu. mm}$ to refer to Tertiary facility
- If Dengue fever is present with other co-morbidities

Management

- Encourage oral fluids. If not tolerated, start intravenous isotonic fluid therapy with or without dextrose at maintenance. Give only isotonic solutions.[*see annexure 1*] Start with 5 ml/kg/hour for 162 hours, then reduce by 2ml/kg/hour every 2 hours till 2ml/kg/hr provided there is clinical improvement and haematocrit is appropriately improving. IV fluids are usually required for 1-2 days.
- Reassess the clinical status and repeat the haematocrit after 2 hours. If the haematocrit remains the same, continue with the same rate for another 264 hours and reassess. If the vital signs/haematocrit is worsening increase the fluid rate and refer immediately.
- Switch to oral as soon as tolerated, total fluid therapy usually 24-48 hrs, titrated to adequate urine output.

Tests for Monitoring:

Frequent recording of vital signs and investigation are essential for evaluating the results of treatment.

- Temperature, Pulse, blood pressure and respiration should be recorded every hour (or more often) until stable subsequently 2 hourly.
- An hourly fluid balance sheet should be kept, recording the type of fluid and the rate and volume of its administration in order to evaluate the adequacy of fluid replacement.
- Chest X-ray, ultrasound abdomen, electrolytes 12-24 hrly as when clinically indicated

Referral Criteria

- All patients with **Warning signs** and signs of **Severe dengue**.
- Patients not clinically responding to therapy in situation.
- Patients with serious co-morbid conditions
- Platelet counts < 50,000/cu.mm with a decreasing trend.

Situation 2

Diagnostic Tests

As in situation 1 in addition the following tests may be useful -

Test	Advantage	Disadvantage
PCR	<ul style="list-style-type: none">• Sensitive• Rapid turn around	<ul style="list-style-type: none">• Expensive
IgM/IgG ratio	<ul style="list-style-type: none">• Can differentiate between primary and secondary dengue	<ul style="list-style-type: none">• No standardisation

Supportive Tests

As in Situation1 and in addition-

- Blood gases.
- Coagulation studies
- Ultrasound
- CT scan
- ECHO

Admission Criteria

Same as in Situation 1 in addition

All patients fulfilling referral criteria form secondary care centres presenting directly or referred.

Pre requisites for management

É Paediatric Intensive care facilities should be available.

Management:

- Obtain a reference haematocrit before fluid therapy. Give only isotonic solutions. Start with 567 ml/kg/hour for 162 hours, then reduce to 365 ml/kg/hr for 264 hours, and then reduce to 263 ml/kg/hr or less according to the clinical response.
- Reassess the clinical status and repeat the haematocrit. If the haematocrit remains the same or rises only minimally, continue with the same rate (263 ml/kg/hr) for another 264 hours. If the vital signs are worsening and haematocrit is rising rapidly, increase the rate to 5610 ml/kg/hour for 162 hours. Reassess the clinical status, repeat the haematocrit and review fluid infusion rates accordingly.
- Give the minimum intravenous fluid volume required to maintain good perfusion and urine output of about 0.5 ml/kg/hr. Intravenous fluids are usually needed for only 24648 hours. Reduce intravenous fluids gradually when the rate of plasma leakage decreases towards the end of the critical phase. This is indicated by urine output and/or oral fluid intake that is/are adequate, or haematocrit decreasing below the baseline value in a stable patient.
- Patients with warning signs should be monitored by health care providers until the period of risk is over.
- Supportive Management- antipyretics, empirical antibiotics as clinically indicated.

Monitoring as follows-

- A detailed fluid balance should be maintained. Parameters that should be monitored include hourly vital signs and peripheral perfusion. (Until the patient is out of the critical phase), urine output (hourly).
- Arterial blood pressure monitoring and central venous pressure monitoring ideal for all children in shock who are fluid unresponsive. Arterial blood gas monitoring as clinically indicated. Infusion pump will help in precise regulation of fluid input.
- Haematocrit (before and after fluid replacement/change, Hourly to 2nd hourly haematocrit for first 6 hours , decreasing frequency as patient improves), Platelet counts 12 hourly blood glucose, and other organ functions (such as renal profile, liver profile, coagulation profile, as indicated).
- Chest x-ray óEffusions, pulmonary edema

- Echocardiogram for assessment of left ventricular function, dimensions and
- regional wall dyskinesia
- ECG to exclude arrhythmia;

Treatment of Shock- See flow chart 1

Treatment of haemorrhagic complications

- A drop in haematocrit with no clinical improvement despite adequate fluid administration indicates significant internal bleeding. Internal bleeding is difficult to recognize in the presence of haemo-concentration. First correct the component of shock according to standard guidelines with early use of packed cell transfusion. Component transfusion is indicated in cases with significant clinical bleeding.
- The results of hematological tests (PT, APTT) may be studied to document the severity of DIC Transfusion of cryoprecipitate and or fresh frozen plasma should be considered in cases of DIC with bleeds.
- Indications for platelet transfusion
 - Shock, acidosis with rapidly declining platelets (greatest risk of DIC)
 - Significant mucosal bleeds (*harbinger of intracranial hemorrhage*)
 - Platelet count < 20,000 cu mm in the acute phase
 - Need for invasive procedures such as central lines maintain platelet count > 50,000 cu mm
 - A low platelet count is less significant after recovery from shock and may not need to be transfused.

Treatment of Fluid overload

Fluid overload with large pleural effusions and ascites is a common cause of acute respiratory distress and failure in severe dengue. Other causes of respiratory distress include acute pulmonary oedema, severe metabolic acidosis from severe shock, and Acute Respiratory Distress Syndrome (ARDS)

Prevention of fluid overload

É When the following signs are present, resuscitation intravenous fluids should be discontinued or reduced to the minimum rate necessary to maintain euglycaemia:

- ó signs of cessation of plasma leakage;
- ó stable blood pressure, pulse and peripheral perfusion;
- ó haematocrit decreases in the presence of a good pulse volume;
- ó afebrile for more than 24-48 days (without the use of antipyretics);
- ó resolving bowel/abdominal symptoms;
- ó improving urine output.

É Aim for a minimum acceptable urine output [0.5ml/kg/hr] to titrate fluids.

É Maintain intravascular volume by using colloids and maintaining oncotic pressure.

The action plan for the treatment of fluid overload is as follows:

- Oxygen therapy/ventilation if indicated should be given immediately.
- Stopping intravenous fluid therapy during the recovery phase will allow fluid in the pleural and peritoneal cavities to return to the intravascular compartment resulting in diuresis.
- Diuretics as given below

If the patient has stable haemodynamic status and is out of the critical phase (more than 24–48 hours of defervescence)

É Stop intravenous fluids but continue close monitoring.

É If necessary, give oral or intravenous furosemide 0.1-0.5 mg/kg/dose once or twice daily, or a continuous infusion of furosemide 0.1 mg/kg/hour. Monitor serum potassium and correct the ensuing hypokalaemia.

If the patient has stable haemodynamic status but is still within the critical phase, reduce the intravenous fluid accordingly.

É Avoid diuretics during the plasma leakage phase.

É Patients who remain in shock with low or normal haematocrit levels but show signs of fluid overload may have occult haemorrhage. Further infusion of large volumes of intravenous fluids will lead only to a poor outcome. If the patient remains in shock and the haematocrit is elevated, repeated small boluses of a colloid solution may help.

Other Complications of Dengue

- Hypo/Hyperglycemia
- Electrolyte abnormalities
- Nosocomial/Co-infection
- Metabolic Acidosis

Should be managed under standard ICU protocols

Supportive Care and Adjuvant Therapy

This may include:

- Renal replacement therapy, with a preference to continuous veno-venous haemodialysis (CVVH), or peritoneal dialysis if the former unavailable;
- Vasopressor and inotropic therapies as temporary measures to prevent life-threatening hypotension in dengue shock and during induction for intubation, while correction of intravascular volume is being vigorously carried out;
- Further treatment of organ impairment, such as severe hepatic involvement or encephalopathy or encephalitis; cardiac abnormalities, such as conduction abnormalities, may occur.

Criteria for discharge:

- Absence of fever for at least 24 hrs.
- Return of appetite.
- Clinical improvement.
- Good urine output.
- Stable haematocrit.
- 2 days after recovery from shock.
- No respiratory distress from pleural effusion and ascitis.

6 Annexure

Immediate replacement of plasma loss/ Issotonic solutions: ^(1,2)

This should be done with any of the following solutions;

- Normal saline.
 - Ringerø lactate
 - In severe/refractory shock, colloids such as Plasma , plasma substitutes (6% hetastarch/dextran/ / 5% albumin /) may be preferred
 - Fresh whole blood or packed red blood cells may be needed for persistent shock despite restoration of fluid volume and a fall in haematocrit, suggesting the possibility of occult blood loss.
- Rapidly administered dextrose containing solution when used for resuscitation may result in hyperglycemia and osmotic diuresis, delaying correction of hypovolaemia. Secondly, dextrose is rapidly metabolized resulting in a hypotonic solution that is inappropriate for shock correction.

Recognition of Shock

The following clinical signs should indicate the presence of shock

Tachycardia , Low pulse volume

Capillary Refill time > 2 sec

Narrow pulse pressure

Blood pressure less than the 3rd centile for age

Cold clammy peripheries

Altered sensorium

Poor urine output [<0.5ml/kg/hr consistently]

Tachypnoea

Metabolic acidosis

Choice of Vasoactive agents/ post resuscitation fluid removal ⁽⁸⁾

- Shock with low BP for age: Dopamine 10mcg/kg/min OR Noradrenaline /adrenaline 0.1-0.2mcg/kg/min
- Shock with normal BP for age: Dobutamine 5-10mcg/kg/min
- Shock with diastolic dysfunction on echo: Milrinone 0.25-0.75mcg/kg/min (no loading dose)
- Predominant pulmonary edema, haemodynamics stable : Nitroglycerine 1-3mcg/kg/min, furosemide infusion 3- 5mg/kg/day, titrate to urine output of 3-5 ml/kg/hr. Cease infusion and infuse fluid if hypoperfusion occurs.
- Pulmonary edema, fluid overload, haemodynamics unstable: Ventilation vital (*high risk of mortality*), can consider peritoneal dialysis if 24 hour experienced nursing and medical staff available in PICU

Good Clinical Practice

- Serial haematocrit measurement (if not bleeding), and urine output provide the most objective guides to fluid replacement and prevention of fluid overload.
- In shock ófluid resuscitate with 10-20ml/Kg of isotonic fluids over 30-60 minutes. Consider in severe shock
- Aim for ≈ 20% fall in haematocrit and adjust fluid rate downwards to avoid overload
- Aim for minimal acceptable urine output (0.5-1ml/kg/hr).
- A urine output > 3 ml /kg /hour indicates Hypervolaemia..
- Fluid replacements are dynamic hence require continuous reassessments.
- No dextrose containing fluid should be used for fluid resuscitation,
- Separate maintenance fluids are usually not required. Glucose/potassium may need to be given separately. Start enteral feeds early.
- All invasive procedures must be performed by most experienced person. If possible, aim for platelets > 50,000/cu mm prior to central line insertion.

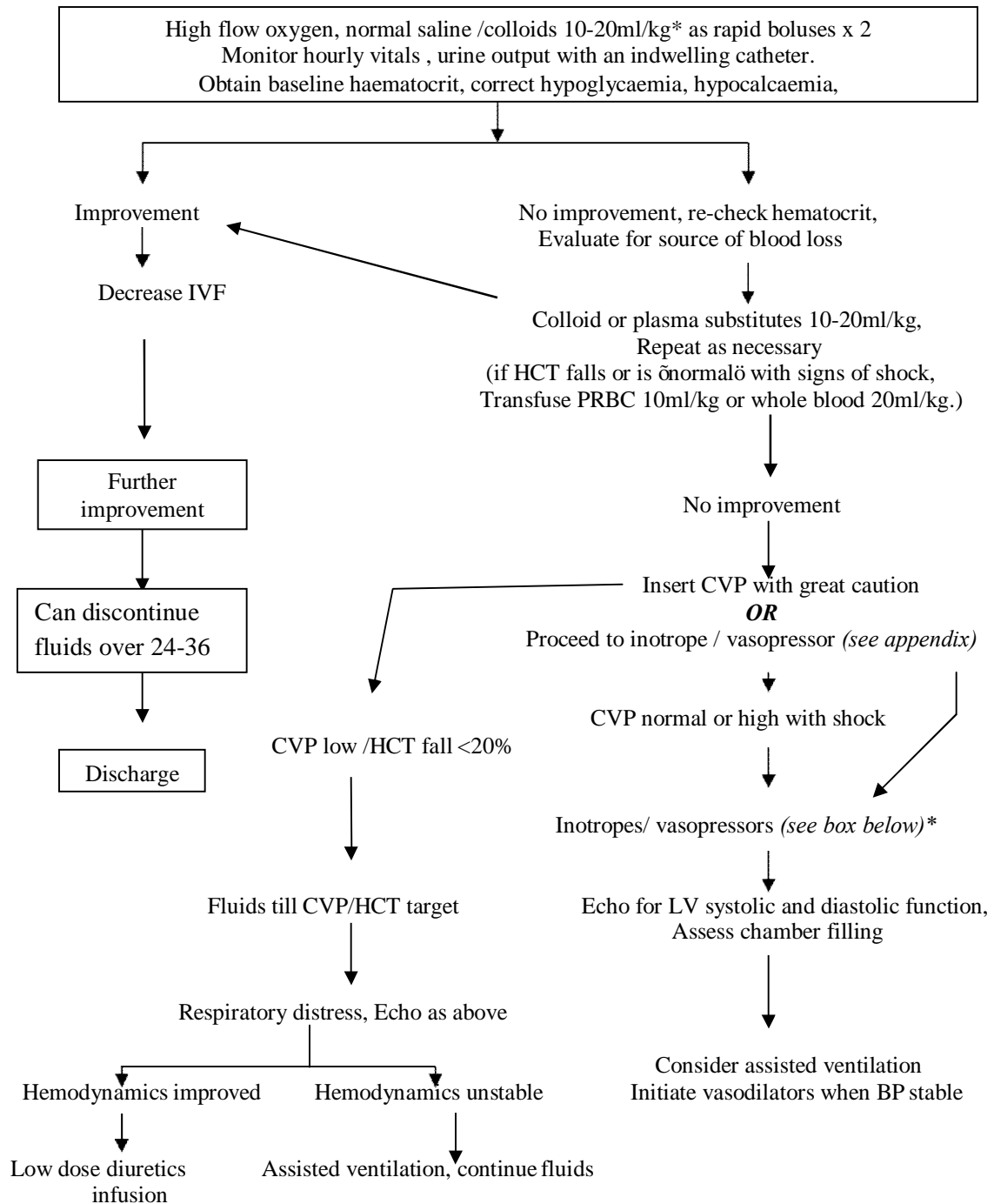
- Profuse bleeds may necessitate transfusion of platelets and FFP regardless of lab values: conversely, low platelet counts in the recovering, stable patient may not be an indication for transfusions.

Flow Chart 1-Volume replacement flow chart for a patient with Severe Dengue and a >20% increase in haematocrit. [No Shock]⁽¹⁾



*- Ideally, infuse replacement and deficit fluid as normal saline/Ringers Lactate (isotonic, non-dextrose containing fluid) and maintenance fluid as ½ DNS with potassium as needed.

Flow Chart 2: Management of Severe Dengue with Shock



References

1. Dengue guidelines for diagnosis, treatment, prevention and control. A joint publication of the World Health Organization (WHO) and the Special Programme for Research and Training in Tropical Diseases (TDR). New Edition 2009
2. Manual of Pediatric emergencies and Critical Care. Dr Suchitra Ranjit. Paras Publications. Second Edition, 2010

EMPYEMA THORACIS

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INTRODUCTION :

Empyema thoracis is a disease of historical importance with modern menace. It results into significant morbidity ; affects precious growing period of a child, parental working days & possible negligence of other family members & also incurs formidable burden on scarce resources if treated improperly or inadequately. Traditionally empyema is being managed empirically. However, with the widespread availability of radioimaging techniques , fibrinolytic agents, safe & effective surgical procedures (open or thoracoscopy) the recent data is leading to more focused management guidelines though optimal management is still controversial (22).

CASE DEFINITION :

Empyema is a term derived from the Greek verb empyein (to suppurate) & literally refers to frank pus in the pleural space. It could be localised or free collection of purulent material in pleural space as a result of combination of inoculation of bacteria & culture medium of pleural fluid. It is an advanced parapneumonic effusion. Pleural space infection is a continuum but classically has been divided into three stages :

Stage 1 or Exudative or Acute phase (lasts upto 3 days) : The inflammatory process associated with the underlying pneumonia leads to accumulation of clear fluid with no or low PMN invasion ,ph >7.3, normal glucose & LDH levels : termed as simple parapneumonic effusion .

Stage 2 or Fibrinopurulent or Transitional phase (3 to 21 days): There is deposition of fibrin in the pleural space leading to septations & formation of loculations. There is increase in white cell count , ph < 7.3, glucose < 40mgs.,LDH >3 times the normal. This is termed as complicated parapneumonic effusion. Eventually when it becomes overt or frank pus which is termed as an empyema. The presence of septations (fibrinous strands

in pleural fluid)doesn't necessarily mean fluid doesn't flow freely, although separate loculations will not communicate with each other.

Stage 3 or Organisational or Chronic phase (> 3 wks . duration): Fibroblasts infiltrate the pleural space & thin intrapleural membranes are reorganised to become thick & non elastic ó the peel or rind. These solid fibrous or leather like peels may prevent lung re-expansion (trapped lung), impair lung function & create a persistent pleural space with potential for infection. At this stage spontaneous healing may occur or a chronic empyema may develop.

Further complications which may occur are : bronchopleural fistula , lung abscess, pneumatocele formation, empyema necessitans : perforation through chest wall, fibrothorax etc

Video-assisted thoracoscopic surgery (VATS) is a key hole or minimal access surgical approach. It achieves debridement of fibrinous pyogenic material, breakdown of loculations, and drainage of pus from the pleural cavity under direct vision. It leaves three small scars.

Mini-thoracotomy achieves debridement and evacuation in a similar manner to VATS but it is an open procedure leaving a small linear scar along the rib line.

Decortication involves an open posterolateral thoracotomy and excision of the thick fibrous pleural rind with evacuation of pyogenic material. It is a longer and more complicated procedure leaving a larger linear scar along the rib line.

INCIDENCE OF CONDITION :

Empyema thoracis constitutes 5-10 % cases seen by a paediatrician in our country(23). The reported rate of empyema thoracis complicating community acquired pneumonia is said to be 27% in children(21). The prevalence of small parapneumonic effusions is difficult to estimate (and often undetected)& they are unlikely to be reported in case series. However cases non secondary to infection viz.. heart disease, malignancy, connective tissue disorders, trauma etc. are largely dependent on the referral base & case mix in the particular hospital (5,6,7,8).

DIFFERENTIAL DIAGNOSIS:

Empyema secondary to bacterial infection eg..
staph./streptococci, Mycobacteria, Anaerobes etc..
Neoplasm
Massive consolidation
Chylous collection
Haemothorax
Lung abscess

PREVENTION & COUNSELING :

Though complete prevention of empyema may not be possible due to factors such as variance in microbial virulence & host's immunological idiosyncracies etc.. but following measures will be very beneficial

* Adequate management of pneumonia ie early recognition, proper selection , duration & mode of delivery of antibiotics according to regional sensitivity pattern & prompt referral facilities to higher centers.

*Prevention of pneumonia :

Ensuring widespread vaccination program for predisposing conditions such as measles, Hib, Pneumococcal, chickenpox etc. because significant fall in incidence of empyema has been reported in vaccinated as compared to non vaccinated zones (21).

Since Staph aureus is the most common organism responsible in our country improving hygienic conditions especially during hot & humid conditions of the year ie April to August will bring down in general incidence & severity of staph infections.

Improvement in nutritional status as PEM is known predisposing factor for all infective illnesses & their complications.

Improvement in dental/oral hygiene as it is a wellknown predisposing factor for development of aspiration pneumonia.

Patients with immunodeficient conditions, tuberculosis, musculoskeletal/neurological illnesses, CHD, Diabetes, Renal disease etc..must remain under high index of suspicion as classical clinical presentation may be absent in such situations.

SITUATION 1 :

At Secondary hospital / Non metro situation : Optimal standards of Treatment in Situations where technology & resources are limited.

*Clinical Diagnosis**

Acute , recurrent or chronic presentations are common.

If a child with pneumonia remains pyrexial or unwell 48 hrs. after the start of the treatment possibility of parapneumonic effusion or empyema should be suspected.

Constitutional signs / symptoms viz.. lassitude, poor appetite, pallor, intermittent fever, easy fatigability, sick look with dull percussion note & decreased breath sounds on auscultation in the setting of partially treated pneumonia, PUO, Disseminated infections e g.. Pyoderma / otitis media /arthritis/Osteomyelitis / Serosal infections etc..

Febrile response may be blunted in immunocompromised patients.

**Physical findings & presentation may vary depending on type organism & duration of illness.*

**Inflammation of pleural space may present with abdominal pain & vomiting.*

INVESTIGATIONS :

- 1) Chest X-ray : Posteroanterior (PA) view
- 2) Ultrasound chest : *Sensitive for confirmation of pleural fluid , for guided diagnostic tapping & insertion of chest drainage tube.*
- 3) Pleural fluid exam:

Colour, Odour, Gram staining, AFB, Bacterial culture, cytology.

Biochemistry: Ph, Sugar, LDH, Proteins.

- 4) *Blood & sputum culture : if feasible*

TREATMENT :

Conservative Management : Antibiotics ± Intercostal Drainage Tube (ICD)

If effusion is simple & small in quantity : can be managed with antibiotics alone .But very close observation is necessary for development of enlarging size &/or compromise of respiratory function when prompt ICD placement is necessary.

Repeated thoracentesis has no role.

If effusion is complicated or frank pus : Antibiotics + ICD

Surgical management : Only if adequately trained personnel & blood banking facilities are available then limited thoracotomy in a situation where there is no satisfactory response (persistent fever, incomplete lung expansion ,loculations on ultrasound etc..)ie.. in stage 2 or fibrinopurulent phase.

Standard operating procedure (SOP)

All the patients of parapneumonic effusion or empyema should be admitted in hospital i e.. no out patient or day care management to be done.

Pediatric surgeon or General surgeon familiar with basic thoracic surgery along with paediatrician or respiratory physician should manage these cases.

They should be monitored closely & carefully by frequent clinical assessment & room air saturation by pulse oximeter whenever child is in resp. distress.

Diagnostic imaging, microbiology, pleural fluid analysis should be carried out promptly.

Conservative management to be started swiftly & supported by antipyretics, analgesia, oxygen ,

if necessary.

Empirical antitubercular therapy should be avoided as far as possible.

Antibiotics :

Intravenous antibiotics for 10 to 14 days for community acquired pneumonia covering Gram positive cocci & anaerobes to be started empirically pending preferably c & s report. Broad spectrum coverage should be started for hospital acquired pneumonia as well as empyema following surgery, trauma & aspiration. Oral antibiotics should be continued at discharge for 1-4 wks. or longer depending on disease state.

Chest drainage tube(ICD) insertion:

Chest drains should be inserted by adequately trained personnel to reduce the risk of complications.

Preferably procedure should be done in operation room or isolated / treatment room on ward. However , if need arises it can be done as a bed side procedure as well.

A suitable assistant and trained nurse must be available.

Routine measurement of the platelet count and clotting studies are only recommended in patients with known risk factors.

Where possible, any coagulopathy or platelet defect should be corrected before chest drain insertion.

Ultrasound should be used to guide thoracocentesis or drain placement.

If general anaesthesia is not being used, intravenous sedation should only be given by those trained in the use of conscious sedation, airway management and resuscitation of children, using full monitoring equipment. .

Local anaesthesia, 2% xylocaine Or .25% bupivacaine, can also be used .

Large bore surgical drains should be inserted at the optimum site suggested by ultrasound, but preferentially placed in the mid axillary line through the ÷safety triangleøø

Substantial force should never be used to insert a drain. Trocar usage preferably should be avoided & should it be needed ,due to circumstances, great care is mandatory to have a guard or control on it while inserting.

Chest tube should be secured well with non absorbable suture & appropriate dressing.

A chest radiograph should be performed after insertion of a chest drain.All chest tubes should be connected to a unidirectional flow drainage system (such as an underwater seal bottle) which must be kept below the level of the patientø chest at all times.

A bubbling chest drain should never be clamped.

A clamped drain should be immediately unclamped and medical advice sought if a patient complains of breathlessness or chest pain.

The drain should be clamped for 1 hour once 10 ml/kg are initially removed.

Patients with chest drains should be managed on wards by staff trained in chest drain management.

When there is a sudden cessation of fluid draining, the drain must be checked for obstruction (blockage or kinking) by flushing.

The drain should be removed once there is clinical resolution.

A drain that cannot be unblocked should be removed and replaced by new catheter if significant pleural fluid remains.

Surgical management :

Proper planning & ensuring availability of all the trained & experienced personnel i.e. surgeon, anaesthesiologist, OT technician & nursing staff and also smooth supply of oxygen, blood, medicines etc. is very important.

Limited thoracotomy with or without rib resection by 5-7cms total incision on either side of chest tube, if already in situ.

To ensure complete lung expansion at the end of the procedure with minimal air leak.

If necrotic lung tissue is present then excision of the segment is to be done.

Send debrided tissue or 'gubbin' for histopathological examination.

ICD removal after complete lung expansion, minimal or no drainage, afebrile state & no air leak

X ray chest to be done before ICD tube removal

Good analgesia (oral &/or suppositories) & early ambulation to hasten the recovery should be practiced regularly.

Antibiotics for 1-2wks. after the discharge are usually sufficient except in situation of complications.

Follow-up :

Till complete resolution of disease process & near complete lung expansion on x-ray chest.

Evaluation of underlying condition, if any.

Referral criteria

If no satisfactory response to conservative management by 5-7 days.

Initial presentation as stage 2 or 3 of an empyema

Suspecting underlying immunodeficiency condition or empyema associated with non pneumonic pathologies which also require specialist's attention.

Development of complications eg Persistent air leak.

Non availability of trained personnel at given time.

Situation 2

At Super Speciality Facility in Metro location where higher end technology is available

Clinical diagnosis*

If a child with pneumonia remains pyrexial or unwell 48 hrs. after the start of the treatment possibility of parapneumonic effusion or empyema should be suspected.

Constitutional s/s viz.. lassitude, poor appetite, pallor, intermittent fever, easy fatigability, sick look with dull percussion note & decreased breath sounds on auscultation in the setting of partially treated pneumonia, PUO, Disseminated infections e g.. Pyoderma / otitis media /arthritis/Osteomyelitis / Serosal infections etc..

Patients inadequately treated or responded to previous therapy.

Complications of an empyema eg.. BPF, lung abscess, empyema necessitans etc.

Patients with underlying conditions such as liver abscess, pancreatitis ,trauma , surgical or endoscopic procedure done etc with respiratory signs & symptoms.

Response may be blunted -absent fever in immunocompromised patients.

**Acute , recurrent or chronic presentations are common.*

**Physical findings & presentation may vary depending on type of organism & duration of illness.*

**Inflammation of pleural space may present with abdominal pain & vomiting*

Investigations

Diagnostic imaging

Posteroanterior or anteroposterior radiographs should be taken; there is no role for a routine lateral radiograph.

Ultrasound may be used to confirm the presence of a pleural fluid collection, septations, to guide thoracentesis or drain placement.

Chest CT scans should not be performed routinely. It should be done once surgery is contemplated to know pleural peel thickness, loculations & their details such as number, position, size etc.; parenchymal pathology, guide for port placement if VATS is being planned .

Diagnostic microbiology

Blood cultures should be performed in all patients with parapneumonic effusion.

When available, sputum should be sent for bacterial culture.

Diagnostic analysis of pleural fluid

Pleural fluid must be sent for microbiological analysis including Gram stain and bacterial culture.

Aspirated pleural fluid should be sent for differential cell count.

Tuberculosis and malignancy must be excluded in the presence of pleural lymphocytosis.

If there is any indication the effusion is not secondary to infection, consider an initial small volume diagnostic tap for cytological analysis, avoiding general anaesthesia/sedation whenever possible.

Biochemical analysis of pleural fluid : Ph, LDH, sugar, & proteins

Diagnostic bronchoscopy

There is no indication for bronchoscopy and it is not routinely recommended. Considered only when bronchoalveolar lavage is necessary or suspected foreign body or assessing bronchial mucosal status for safe closure of br. stump when major pulmonary resection is also planned alongwith decortication .

Treatment

Conservative management (antibiotics ± simple drainage)

Effusions which are enlarging and/or compromising respiratory function should not be managed by antibiotics alone.

Give consideration to early active treatment as conservative treatment results in prolonged duration of illness and hospital stay.

If a child has significant pleural infection, a drain should be inserted at the outset and repeated taps are not recommended.

Antibiotics

All cases should be treated with intravenous antibiotics and must include cover for Gram positive cocci eg..Staph Aureous, Streptococci & Anarobes.

Broader spectrum cover is required for hospital acquired infections, as well as those secondary to surgery, trauma, and aspiration.

Where possible, antibiotic choice should be guided by microbiology results.

Oral antibiotics should be given at discharge for 164 weeks, but longer if there is residual disease.

Chest drains

Chest drains should be inserted by adequately trained personnel to reduce the risk of complications.

A suitable assistant and trained nurse must be available.

Routine measurement of the platelet count and clotting studies are only recommended in patients with known risk factors.

Where possible, any coagulopathy or platelet defect should be corrected before chest drain insertion.

Ultrasound should be used to guide thoracocentesis or drain placement, when available.

If general anaesthesia is not being used, intravenous sedation should only be given by those trained in the use of

conscious sedation, airway management and resuscitation of children, using full monitoring equipment.

Large bore surgical drains should be inserted at the optimum site suggested by ultrasound. The usual site for ICD insertion should be in the mid axillary line in the 5th intercostal space which is in the "safe triangle".

Substantial force should never be used to insert a drain. Trocar usage preferably should be avoided & should it be needed, due to circumstances, great care is mandatory to have a guard or control on it while inserting.

Chest radiograph should be performed after insertion of a chest drain.

All chest tubes should be connected to a unidirectional flow drainage system (such as an underwater seal bottle) which must be kept below the level of the patient's chest at all times.

Appropriately trained nursing staff must supervise the use of chest drain suction.

A bubbling chest drain should never be clamped.

A clamped drain should be immediately unclamped and medical advice sought if a patient complains of breathlessness or chest pain.

Patients with chest drains should be managed on specialist wards by staff trained in chest drain management.

When there is a sudden cessation of fluid draining, the drain must be checked for obstruction (blockage or kinking) by milking / flushing. If it can not be unblocked in presence of significant pleural infection then it should be reinserted.

The drain should be removed once there is clinical resolution & / or lung expansion on x-ray.

Intrapleural fibrinolytics

Intrapleural fibrinolytics are said to shorten hospital stay and may be used for any stage 2 empyema .

There is no evidence that any of the *three* fibrinolytics (Streptokinase, Urokinase, Alteplase) are more effective than the others, but only urokinase has been studied in a randomised controlled trial .

Urokinase should be given twice daily for 3 days (6 doses in total) using 40 000 units in 40 ml 0.9% saline for children weighing 10 kg or above, and 10 000 units in 10 ml 0.9% saline for children weighing under 10 kg.

Surgery

Patients should be considered for surgical treatment if they have persisting sepsis in association with a persistent pleural collection, despite chest tube drainage and antibiotics.

Failure of chest tube drainage, antibiotics, and fibrinolytics would necessitate surgical intervention. However, a pediatric surgeon should be involved early in the management of empyema thoracis.

If requisite skill & facilities are available then VATS debridement is preferable over open surgical procedure in stage 2 & select stage 3 empyema cases.

Organised empyema in a symptomatic child may require formal thoracotomy and decortication.

Other management

Antipyretics should be given.

Analgesia is important to keep the child comfortable, particularly in the presence of a chest drain.

Early mobilisation, chest physiotherapy and exercise is recommended.

Secondary scoliosis noted on the chest radiograph is common but transient; no specific treatment is required but resolution must be confirmed.

Standard operating procedure (SOP)

All the patients of parapneumonic effusion or empyema should be admitted in hospital i.e. no out patient or day care management to be done.

Pediatric surgeon or a surgeon well trained in pediatric thoracic surgery along with paediatrician or respiratory physician should manage these cases.

CECT(Contrast Enhanced Cat Scan) should be done if surgery is contemplated.

There are no evidence based criteria to guide the decision on when a child should proceed to surgery, and consequently there is little consensus on the role of medical versus surgical management (1)

Intrapleural fibrinolytics

Intrapleural fibrinolytics shorten hospital stay and are recommended for any complicated parapneumonic effusion (thick fluid with loculations) or empyema (overt pus).

There is no evidence that any of the three fibrinolytics (streptokinase, urokinase & tPA) are more effective than the others, but only urokinase has been studied in a randomised controlled trial in children so is recommended (10 to 16)

Surgery:

The decision to involve a pediatric surgeon early in the decision making process should be encouraged and referral should not automatically mean surgery is inevitable. (1)

Available procedures are: VATS, Mini thoracotomy & Decortication.

A chest drain(s) is left after each procedure for further drainage of fluid/pus.

A persistent radiological abnormality in a symptom-free well child is not an indication for surgery.

Role of surgical management in complex empyema

(A) Organised empyema with a thick fibrous peel

Organised empyema in a symptomatic child may require formal thoracotomy and decortication.

The surgical management of an organised empyema, in which a thick fibrous peel is restricting lung expansion and causing chronic sepsis with fever, requires a formal thoracotomy with excision of the pleural rinds (decortication) to achieve proper lung re-expansion. However, if the child is asymptomatic, surgery is not necessarily indicated. (18)

(C) Bronchopleural fistula and pyopneumothorax

Different approaches have been advocated for a bronchopleural fistula related to an empyema. Most fistulae are peripheral and the majority resolve with continued chest drainage and antibiotics provided the lung shows satisfactory lung expansion.

However, at times they are slow and difficult to resolve, and it has been said that conservative management and open thoracostomies result in protracted recovery and morbidity. A more radical approach is partial decortication and muscle flap surgery to bring a blood supply to the necrotic area and help with healing the fistula. This can either be done as a staged procedure or a more aggressive one stage approach (19, 20)

Follow up

Children should be followed up after discharge until they have recovered completely and their chest radiograph has returned to near normal.

Underlying diagnosis – for example, immunodeficiency – may need to be considered in selected situations.

open chest tube care in select patients till tube is in situ

For monitoring of ATT

Referral to tertiary speciality centre

Non availability of skilled & experienced personnel or infrastructure necessary for management of complex situations eg. BPF, major lung resections or Thoracoscopic procedure or need of PICU facilities etc..

When respiratory paediatrician is needed to be involved early in the care of patients requiring chest tube drainage for a pleural infection & in critically ill child.

Who does what ? & Time lines :

Doctor : Pediatric Surgeon :

Does all the interventional procedures & involved in decision making regarding overall management with paediatrician.

Pediatrician (Respiratory) : Along with surgeon involved in conservative management & post operative care & preop. preparation

General Surgeon : Does interventions In situation 1 only & in situation 2 maybe involved in assisting pediatric surgeon.

Anaesthesiologist : Anaesthesia & pain management

Junior doctor : Assisting in surgery, ICD care ,dressings & day to day ward management

b) OT Technician : Helping anaesthesiologist & surgeon in OT & if required on ward bedside interventions.

C) Nursing Staff : Care & organisation of instrument trolley, medications & ICD care, suction etc. & day to day nursing care.

Time line :

Radiological clearance takes few wks. to many months.

If an empyema fails to respond in 3-5 days of conservative treatment then surgical intervention should be considered to reduce morbidity.

Dwelling time for fibrinolytics is 1-4 hours.

There is no specific / precise time period for spontaneous closure or need for surgical intervention in BPF patients.

Antibiotics are needed for 4-8 wks. (Intravenous 2-3 wks. & oral 1-4wks.)

ATT is required for 6-9 mths duration.

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Resources required for one patient / procedure

Situation 1

Human resources : Surgeon 1, Pediatrician1, nursing staff1, OT technician 1, Anaesthesiologist

Investigations :

Chest radiograph

Ultrasound scan of chest

Full blood count

C-reactive protein (some regard this as a useful marker of progress)

.Blood culture (including anaerobic bottle) & Sputum culture (if available)

Drugs & Cosumables :

Antibiotics

,Analgesics,antipyretics,Oxygen,IVFluids,sedatives,atropine/pyrolate,ketamine, propafol,local anaesthetics,muscle relaxants,antiemetics, PPI/H2 blockers,Emergency medicationsetcí ,Betadine, Savolon, spirit, Normal saline Syringes & needles,IVsets,oxygen mask,IVCannulae,Sticking plaster,Cotton, gauze pieces, pads,suture material,ICD tubes, Under water seal drainage bags etcí Gloves (surgical),Gowns, Eye towel

Equipments :

EQUIPMENT FOR CHEST DRAIN INSERTION

Sterile gloves and gown

Skin antiseptic solution, e.g. povidone iodine (Betadine) or

chlorhexidine in alcohol

Sterile gauze swabs

A selection of syringes (2 ml and 5 ml) and needles (21&25gauge)

Local anaesthetic, e.g. 0.25% bupivacaine (Marcaine) , 2% xylocaine

Scalpel and blade

Suture (e.g. 2/0 or 3/0 silk)

Guide wire with dilators for Seldinger technique

Chest tube: 10&12 FG appropriate for most children (8&14 FG should be available)

Connecting tubing

Closed drainage system (including sterile water if underwater seal being used)

Sterile universal containers and anaerobic blood culture bottle for pleural fluid

Large transparent adhesive dressings

Equipment for percutaneous long line and bottles for blood tests.

Limited Thoracotomy Equipments :

Basic Thoracotomy set

General Anaesthesia machine, Tracheal intubation set organised well in operation room with other basic facilities eg. Good negative suction source, satisfactory illumination, patient warming gadgets etc..

Situation 2

Human resources : Well trained pediatric surgeon, Respiratory pediatrician & Anaesthesiologist mandatory ,Assistent Surgeon, Resident doctors & specialised nursing staff round the clock, Anaesthesia technician.

Investigations : As in situation 1 + Albumin, Creatinine, blood group ,Blood gases, Specialised infective, immunodeficiency etcí workup facilities + Contrast Enhanced CT Scan + investigations facilities for associated problems & complications from the disease process.

Equipments :

Basic + High end open surgery & Thoracoscopy set in well planned & equipped OT.

Drugs & Consumables

INGUINAL HERNIA IN CHILDREN

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a) WHEN TO SUSPECT/ RECOGNIZE?

Inguinal hernia is suspected in any child with a swelling in the inguinoscrotal region.

a. Introduction:

Inguinal hernia repair is one of the most common pediatric operations performed. Most hernias that present at birth or in childhood are indirect inguinal hernias. All pediatric inguinal hernias require operative treatment to prevent the development of complications, such as inguinal hernia incarceration or strangulation.

b. Case definition:

Inguinal hernia is a type of ventral hernia that occurs when an intra-abdominal structures, such as bowel or omentum, protrude through the open processus vaginalis through the inguinal canal.

b) INCIDENCE OF THE CONDITION IN OUR COUNTRY

Although the exact incidence of indirect inguinal hernia in infants and children is unknown, the reported incidence ranges from 1-5%. Sixty percent of hernias occur on the right side. Premature infants are at increased risk for inguinal hernia, with incidence rates of 2% in females and 7-30% in males.

Inguinal hernias are much more common in males than in females. The male-to-female ratio is estimated to be 4-8:1.

Premature infants are at an increased risk for inguinal hernia, with the incidence ranging from 7-30%. Moreover, the associated risk of incarceration is more than 60% in this population.

c) **DIFFERENTIAL DIAGNOSIS**

Congenital Hydrocoele

Inguinal adenitis

Femoral adenitis

Psoas abscess

Saphenous varix

Retractile testis

Varicocele

Testicular tumor

Undescended testis

d) **PREVENTION AND COUNSELING :**

A high index of suspicion is required-especially in the high risk population of premature babies. If a child has developed a unilateral hernia, there is a potential risk of developing a hernia on the opposite side-this risk is higher in premature babies and infant girls. These families need to be counseled about signs and symptoms of these recurrences.

e) **OPTIMAL DIAGNOSTIC CRITERIA, INVESTIGATIONS, TREATMENT & REFERRAL CRITERIA**

Diagnostic criteria: diagnosis is clinical.

1. History of a soft swelling in inguinal region. Gets larger when child cries, may disappear completely when the child is quietly lying down.
2. Examination: Soft, reducible mass in the inguinal area is diagnostic. Even in the absence of the mass at examination, a strong history is adequate for diagnosis.

Investigations:

No imaging studies are required. General tests towards anaesthesia fitness may be required (haemoglobin, urine analysis).

Referral Criteria:

A strong clinical history and physical findings of inguinal hernia are indications for referral for surgery.

HISTORY

The child with an inguinal hernia presents with a bulge at the internal or external ring or within the scrotum. The parents typically provide the history of a visible swelling or bulge, commonly intermittent, in the inguinoscrotal region in boys and inguinolabial region in girls.

Usually, a simple inguinal hernia in an infant is painless.

The bulge commonly occurs after crying or straining and often resolves during the night while the baby is sleeping.

If the family provides a history of a painful bulge in the inguinal region, one must suspect the presence of an incarcerated inguinal hernia. Patients with an incarcerated hernia generally present with a tender firm mass in the inguinal canal or scrotum. The child may be fussy, unwilling to feed, and inconsolably crying. The skin overlying the bulge may be edematous, erythematous, and discolored.

EXAMINATION

Physical examination of a child with an inguinal hernia typically reveals a palpable smooth mass originating from the external ring lateral to the pubic tubercle. The mass may only be noticeable after coughing or performing a Valsalva maneuver and it should be reduced easily. Occasionally, the examining physician may feel the loops of intestine within the hernia sac. In girls, feeling the ovary in the hernia sac is not unusual; it is not infrequently confused with a lymph node in the groin region. In boys, palpation of both testicles is important to rule out an undescended or retractile testicle.

Hernia and hydrocele: Transillumination has been advocated as a means of distinguishing between the presence of a sac filled with fluid in the scrotum (hydrocele) and the presence of bowel in the scrotal sac. However, in cases of inguinal hernia incarceration, transillumination may not be beneficial because any viscera that are distended and fluid-filled in the scrotum of a young infant may also transilluminate.

INVESTIGATIONS

No laboratory studies are needed in the assessment of a patient with a suspected inguinal hernia and/or hydrocele.

Ultrasonography: Its routine use is unnecessary. It is indicated when presentation and examination suggest a diagnosis other than hernia or hydrocele. An enlarged inguinal lymph node can mimic an incarcerated inguinal hernia.

Laparoscopy: Diagnostic laparoscopy may rarely be required for determining the presence of an inguinal hernia. It is used only in the following: a) assessment of contralateral hernia when one is being operated upon, and b) recurrent hernia after previous surgery.

TREATMENT

Congenital hernias are treated surgically with herniotomy. Surgical treatment can be either open or laparoscopic. Inguinal hernias do not spontaneously heal and must be surgically repaired because of the ever-present risk of incarceration. Repair is usually planned as an elective procedure as soon as possible after diagnosis.

If hernia is irreducible, ie cannot be easily pushed back into the abdomen, child needs to be admitted and a manual reduction tried by an experienced pediatric surgeon. If successful, the operation is performed after 24-48 hours to allow local oedema to settle down.

If reduction is unsuccessful, or if there is clinical evidence of inflammation (as evidenced by pain, redness, edema of skin on hernia) emergency exploration and hernia repair is necessary.

Hydroceles without hernia in neonates: This is the only exception in which surgical treatment may be delayed. Repair of hydroceles in neonates without the presence of hernia is typically delayed for 12 months because the connection with the peritoneal cavity (via the processus vaginalis) may be very small and may have already closed or be in the process of closing. If the hydrocoele persists after this observation period, operative repair is indicated and appropriate.

Postpone the operation in the event of upper respiratory tract infection, otitis media, or significant rash in the groin.

FOLLOW UP

No specific limitations are indicated once the diagnosis of an inguinal hernia has been established; however, following operative repair, avoidance of major physical activity for 1 week is recommended. After that time, the patient is allowed to participate in physical activities (eg, sports, swimming, running).

Children younger than 5 years are likely to recover extremely quickly from surgery; they are typically capable of returning to their normal level of activities within 24-48 hours of surgery.

*Situation 1: At Secondary Hospital/ Non-Metro situation: Optimal Standards of Treatment in Situations where technology and resources are limited

a. Clinical Diagnosis:

Similar generic diagnostic criteria apply. A typical history and physical finding are adequate for diagnosis.

- b. **Investigations:**
 - a. Investigations are not required other than for anaesthetic purposes.
 - b. Rarely, ultrasonography and diagnostic laparoscopy may be indicated

- c. **Treatment:**
 - a. Inguinal hernia: Treatment is surgery -herniotomy operation that aims at ligating the patent processus vaginalis at the internal ring after reduction of contents into the abdomen
 - b. Congenital hydrocoele: Treatment is deferred until the 2nd birthday as there is a 80% chance of spontaneous closure. Surgery is indicated if hydrocoele persists beyond the 2nd year and if it is rapidly growing in size.
 - c. All hernia and hydrocoele repairs in infants and children MUST be performed by a qualified pediatric surgeon.

Standard Operating procedure

- a. In Patient : inpatient care is indicated if:
 - I. h/o incarceration or obstructions
 - II. neonate awaiting hernia repair
- b. Out Patient
 - i. Outpatient care is adequate for diagnosis and follow up in most children
 - ii. Clinical evaluation usually sufficient to diagnose
- c. Day Care
 - i. most hernia operations in children are done as day care procedures

d. Referral criteria:

A child with a hernia needs referral to a higher centre if:

1. neonatal age and anaesthesia facilities are inadequate
2. ex-premature baby who had prolonged ventilation
3. recurrent inguinal hernia
4. inguinal hernia with complications (incarceration, obstruction)
5. associated major morbidity-such as cardiac anomalies, lung disease, renal disease, ascites

***Situation 2: At Super Specialty Facility in Metro location where higher-end technology is available**

α. **Clinical Diagnosis:**

α. Same as in situation1

β. **Investigations:**

α. Same as in situation1

β. additional tests for comorbid conditions such as pulmonary and cardiac anomalies

χ. **Treatment:**

a. principles of treatment are same as above

Standard Operating procedure

α. In Patient

i. criteria same as above

ii. all babies under 1 year of age need inpatient care after surgery-as there is an increased risk of apnoea in this subgroup.

iii. All children with comorbid problems-cardiac, respiratory or others, need inpatient care after surgery

b Out Patient

i. criteria same as above

c. Day Care

ii. criteria same as above

iii. Children who are above 1 year of age and in good health with no associated comorbid problems can be treated as day care procedures

δ. **Referral criteria:**

No further referrals

f ***WHO DOES WHAT? and TIMELINES***

a. Doctor makes a clinical diagnosis, counsels the family and plans surgery- a pediatric surgeon performs the surgery

b. Nurse: assists surgeon in care of child during pre, intra and post operative course of the baby

c. Technician: assists medical and nursing teams in care of child during intra and post-operative periods.

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RESOURCES REQUIRED FOR ONE PATIENT / PROCEDURE (PATIENT WEIGHT 60 KGS)

(Units to be specified for human resources, investigations, drugs and consumables and equipment. Quantity to also be specified)

Situation	Human Resources	Drugs & Consumables	Equipment
1	<ul style="list-style-type: none"> • Pediatric Surgeon • Pediatrician • Pediatric Nurse • Lab. Technician 	<ul style="list-style-type: none"> • I.V. Glucose/ Fluids • I.V. cannula • I.V. Set • anesthetic drugs, disposables • antibiotic prophylaxis 	<ul style="list-style-type: none"> • Radiant Warmer • Saturation monitor • Basic Lab • Child friendly OT
2	<ul style="list-style-type: none"> • Pediatric surgeon • Pediatrician • Pediatric anaesthetist • Pediatric Nurse 	<ul style="list-style-type: none"> • I.V. Glucose/ Fluids • I.V. cannula • I.V. Set • anesthetic drugs, disposables • antibiotic prophylaxis 	<ul style="list-style-type: none"> • ICU • Pediatric O.T.

NEONATAL CHOLESTASIS

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I. WHEN TO SUSPECT/ RECOGNIZE?

Introduction:

Neonatal cholestasis is a pathological condition in the newborn where in bile flow from the liver is reduced. Neonatal Cholestasis Syndrome (NCS) includes a wide spectrum of clinical conditions ranging from congenital malformations of the hepatobiliary tree, infections, inborn errors of metabolism to some of the recently identified clinical conditions with or without genetic predilection. Most of these disorders have linkage with insults during antenatal, natal and postnatal periods.

NCS has largely remained ignored in our country.

Case definition:

Neonatal cholestasis refers to conjugated hyperbilirubinemia $>1.5 \text{ ó } 2 \text{ mg\%}$ and/or direct component of more than 20% of total bilirubin in a newborn/ infant with passage of high coloured urine with or without clay stools.

II. INCIDENCE OF THE CONDITION IN OUR COUNTRY

NCS constitutes 30% of referrals with hepatobiliary disorders in India. The average age of presentation to a specialized center is 3.5 months (range birth to 15 months) with a consequent delay of 3 months in referral (medical and surgical centers).

Based on consensus conference by paediatric gastroenterology, out of 1008 cases analysed in our country

- Hepatocellular causes: 53 % (neonatal hepatitis-47%, metabolic-4%, others-2%)
- Obstructive causes: 38 % (biliary atresia-34%, Choledochal cyst-4%)
- Ductal paucity: 3%
- Idiopathic: 6%

In neonatal hepatitis-

- Idiopathic giant cell hepatitis: 64%,
- TORCH: 22%
- Sepsis: 8%
- Others: 6%

III. DIFFERENTIAL DIAGNOSIS

A. CAUSES OF EXTRA HEPATIC OBSTRUCTION

- Biliary Atresia
- Choledochal Cyst
- Spontaneous perforation of bile ducts
- Biliary stenosis
- Inspissation of bile ducts
- Mass/peritoneal bands

B. HEPATOCELLULAR

1. INFECTIVE

- Sepsis
- TORCH
- Malaria
- UTI
- Hepatitis
- Other Viral infections
- HIV

2. METABOLIC

- Galactossemia
- Hereditary Fructosemia
- Tyrosinemia
- Alfa 1 AT deficiency
- Bile acid disorders
- Fatty Acid Oxidation defects
- Cystic fibrosis
- Storage disorders
- Neonatal hemochromatosis
- Zellweger's disease

3. MISCELLANEOUS

- TPN
- Shock
- Hypoperfusion
- Downs
- Congenital heart / valvular abn

4. IDIOPATHIC

C. PAUCITY OF INTRAHEPATIC DUCTS

- **Syndromic** - Alagille's syndrome, Byler's, Aagene's
- **Non - Syndromic** - a 1AT deficiency, Idiopathic, Familial

IV. PREVENTION AND COUNSELING

A high index of suspicion is necessary. Mothers must be informed about the need to seek medical attention if jaundice persists beyond two weeks of birth and / or baby passes pale stools and high coloured urine. If the previous sibling has had liver disease antenatal counselling and referral for further evaluation may be necessary.

V. OPTIMAL DIAGNOSTIC CRITERIA, INVESTIGATIONS, TREATMENT & REFERRAL CRITERIA

a. Diagnostic criteria:

1. Clinical: Neonate with jaundice persistent beyond 2 weeks, dark colour urine and/or pale stool
2. Screening Biochemistry: Serum bilirubin direct and indirect

Any child that meets with the clinical and /or biochemical criteria needs investigation, treatment and referral.

Typical presentation:

- Newborn with jaundice/ high colored urine with or without clay colour stools beyond two weeks of age.
- Typically a child with biliary atresia is usually a term baby with normal weight, accepting feeds well.
- Pigmented stools do not rule out Biliary atresia, upto 30% of biliary atresia stools are yellow in the early weeks.

Clinical examinations:

Clinical evaluation-

1. Sick baby: Sepsis/UTI/Congenital infections/metabolic disorders.
2. Dysmorphism : Down's syndrome/ alagilles syndrome.
3. Examination of eye and fundus must be done-
 - Cataract in Galactosemia
 - Chorioretinitis in TORCH
 - Posterior embryotoxins in alagilles syndrome
 - Cherry red spot in Lipid storage disorders
4. Chronic Cholestasis – Pruritis/ irritability/xanthomas
5. Failure to thrive.

***Situation 1: At Secondary Hospital/ Non-Metro situation: Optimal Standards of Treatment in Situations where technology and resources are limited**

- e. **Clinical Diagnosis:**
2. Jaundiced child
 3. Dark urine
 4. Pale stool
 5. Hepatomegaly \pm splenomegaly
- f. **Investigations:**
- Hematology: CBC
 - Urine routine & microscopy
 - Biochemistry:
 - a. LFT
 - b. RFT
 - c. PT/INR
 - d. RBS
 - Imaging: USG Abdomen
- g. **Treatment:** _____

Standard Operating procedure

4. Resuscitation if required,
 5. Correction of Hypoglycemia
 6. Administration of Vitamin K (0.3mg/kg parenteral)
 7. Initiation of antibiotics: if there is clinical or laboratory evidence of infection or sepsis
- a. **In Patient** : Child needs admission if
1. there is clinical evidence of dehydration
 2. clinical \pm laboratory evidence of hypoglycemia
sepsis or coagulopathy.
 3. failure to thrive
- b. **Out Patient** : Baby who is clinically well, feeding well and has no evidence of hypoglycemia or coagulopathy can be investigated as an outpatient

c. **Day Care** : No role of day care admission.

h. **Referral criteria:** (All workup at this level must be completed by 48 hours)

1. Any child with neonatal cholestasis syndrome who is > 2 weeks of age
2. Clinically unwell, poor feeding, poor weight gain
3. Evidence of coagulopathy, hypoglycemia or sepsis

***Situation 2: At Super Specialty Facility in Metro location where higher-end technology is available**

Clinical Diagnosis:

Investigations: Urgent Investigations

- Blood counts
- LFT
- PT
- Electrolytes
- Blood culture
- Urine culture, Urine microscopy
- Urine reducing substances
- GRBS
- Ascitic tap (if ascites)

Standard LFT are usually abnormal with modestly raised levels of AST, ALT, and alkaline phosphatase. GGT is raised in all cases of cholestasis except in one of the bile acid synthetic defects. Serum albumin does not fall till late. None of the biochemical tests are of deciding value and at best reflect the degree of damage to liver.

Tests directed towards infective and metabolic causes :

Blood Tests

- TORCH, VDRL, Hepatitis B/C, HIV
- T4, TSH
- Serum cortisol
- α 1 AT levels and phenotype
- Galactose 1 Phosphate Uridyl transferase (to r/o galactosemia)
- Urinary succinyl acetone (to r/o tyrosinemia)
- Cholesterol, triglycerides
- S. iron and ferritin levels (to r/o neonatal hemochromatosis)

Radiology

Role of USG

USG can exclude choledochal cyst, any focal lesions, dilated CBD, anomalies of viscera or portal hypertension.

Role of Hida Scan

Hepatobiliary scintigraphy, after a 5 day priming with phenobarbitone, is useful. Excretion of the radio-tracer into the gut rules out biliary atresia. However, the converse is not true and absence of gut excretion of radiotracer requires further evaluation.

Role of Liver Biopsy

Liver biopsy is useful in the characterisation of NCS in some cases. Coagulopathy and ascitis are contraindications for percutaneous liver biopsy.

TREATMENT

On suspicion of cholestatic liver disease, vitamin-K is started along with supplementation of other fat soluble vitamins (A,D,E,)

Treatable Causes

Medical

- Sepsis
- UTI
- Congenital infections
- Hepatitis
- Galactosemia
- Hereditary fructose intolerance
- Hypothyroidism
- Hypopituitarism
- Tyrosinaemia

Surgical

3. Biliary Atresia
4. Choledochal cyst
5. Spontaneous perforation of bile ducts
6. Inspissation of bile ducts

SUPPORTIVE CARE

Nutrition

Energy - 125% RDA

Protein intake -2-3 gm/kg/day in infants (0.5gm/kg/day in hepatic encephalopathy)

Vitamin A should be supplemented in a dose of 50,000 IU intramuscularly at diagnosis and then 10,000 IU monthly till cholestasis resolves. Avoid hyper- vitaminosis as it can enhance fibrosis.

Vitamin D should be supplemented in a dose of 30,000 IU intramuscular at diagnosis and then monthly till cholestasis resolves. If the child has rickets give a dose of 60,000 IU.

Oral Vitamin E, supplementation (50-200 mg/day) is required to avoid neuro-muscular degeneration, retinal pigmentation and hemolytic anemia.

Provide Vitamin K 5 mg/day intramuscular/intravenous for first 3 days and then 5 mg weekly. Perform prothombin time (PT) monthly. Administer injectable vitamin K if PT is prolonged.

Water soluble vitamins and trace elements (2-5) times RDA)

Pruritis

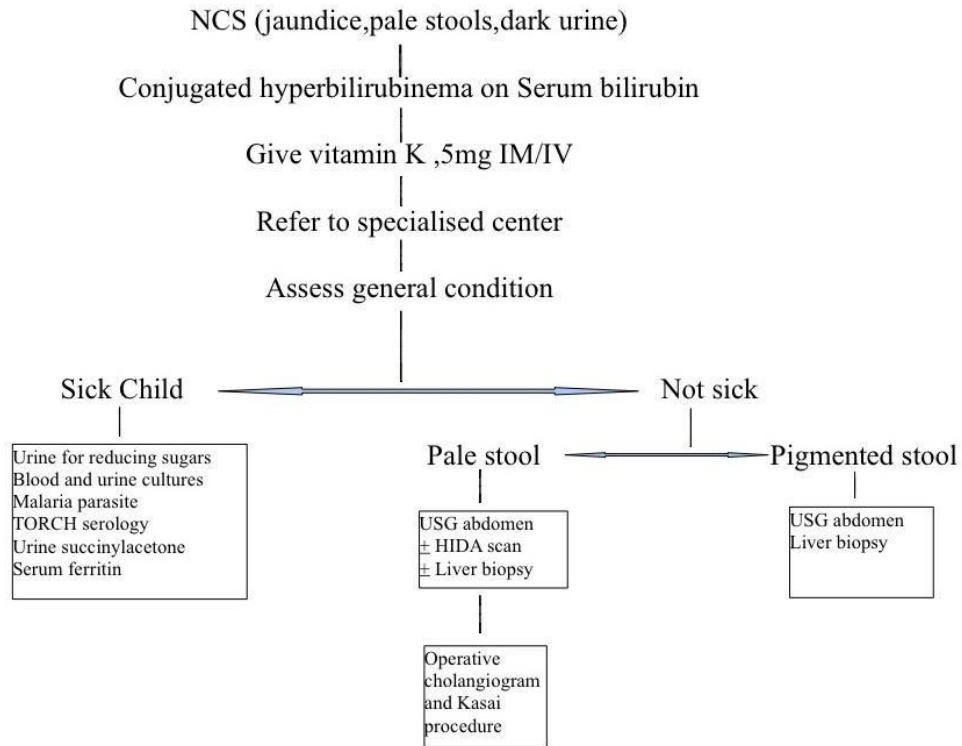
For control of pruritis following agents have been tried :-

1. Phenobarbitone-5mg/kg/day
2. Rifampicin- 10mg/kg/day
3. Ursodeoxycholic acid-10-20 mg/kg/day
4. Cholestyramine-4- 8gm/kg/day
5. Terfenadine 1-3mg/kg/day
6. Carbamazepine

Liver Transplantation

This may remain the only option for infants with decompensated liver disease (ascites and /or encephalopathy) or failed portoenterostomy.

TREATMENT PLAN



Standard Operating procedure

In Patient only: May require ICU monitoring

Referral criteria:

Referral criteria for a specialist centre:

Any case of neonatal cholestasis as defined by above parameters with deranged liver function tests to be referred to tertiary centre for further management.

Child needs to be referred to a specialist pediatric liver unit if,

1. Evidence of progressive liver failure
2. Evidence of complications such as portal hypertension, SBP, Respiratory distress, pathological fractures.
3. Failure of Kasai operation
 - Evidence of liver cirrhosis in biopsy
 - Jaundice not cleared by 2 months after surgery
4. Considerations for liver transplant

Situation 2:

Referred cases from secondary centres or any newborn with evidence of cholestasis with deranged liver function tests.

Investigation:

Medical causes	Surgical causes
<p>As in situation 1 & Additional investigations</p> <p>Blood culture Urine culture Urine reducing substances Prothrombin time T3/T4/TSH TORCH/VDRL Hep B/C, HIV Metabolic work up</p>	<p>USG Abdomen:</p> <p>Day 5 : Prepare patient for HIDA (Priming with gardenal 3-5 mg/kg)</p>

Subsequent investigation (if initial workup negative)

- α 1 antitrypsin levels & phenotype
Galactose 1- P uridyl transferase
Urinary succinyl acetone
Cholesterol, Triglycerides
Serum ferritin, iron
Radiology
Rare investigation-

Bone marrow aspiration/skin biopsy/muscle biopsy/ serum lactate / pyruvate / ammonia, very long chain fatty acid, urinary organic acids, urinary bile acids, auto immune screen / sweat chloride test.

Treatment:

- Aims of treatment
Within 7 days of hospitalisation treatment is mandatory.
- Kasai's Portoenterostomy preferably within 60 days of life.

Medical:

Neonatal hepatitis:

2. Bacterial Sepsis/ UTI : antibiotics
3. Malaria : antimalarial
4. Toxoplasmosis & syphilis : specific antibiotics
5. CMV : ganciclovir
6. Herpes : acyclovir
7. *Metabolic:*
 8. Galactosemia : stop lactose milk
 9. Fructosemia : withdrawal of fructose containing item
10. Hypothyroidism : Thyroxine

Chronic cholestatis:

Basically improve nutritional status

Provide energy (125%RDA)
 Proteins intake 2-3 gm/kg
 MCT diet
 Fat Soluble vitamins (A/D/E/K)
 Pruritis control:
 Phenobarbitone / Rifampicin/ UDCA/ Cholestyramine/ Terfenadine/ Carbamazepine

Surgical

Biliary atresia	Kasai Portoenterostomy
Choledochal cyst	Laparatomy
Failed Kasai End stage liver disease PFIC	Liver Transplantation

VI. WHO DOES WHAT? and TIMELINES

- **Doctor :**
 - Close and continuous and crucial monitoring of the child
 - Planning of tests and treatment
 -
- **Nurse**
 - Counseling and support to child and family
 - Assisting to treatment
- **Technician**

VII. FURTHER READING / REFERENCES

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RESOURCES REQUIRED FOR ONE PATIENT / PROCEDURE (PATIENT WEIGHT 60 KGS)

(Units to be specified for human resources, investigations, drugs and consumables and equipment. Quantity to also be specified)

Situation	Human resources	Investigations	Drugs & consumables	Equipment
1	Pediatrician Pediatric Nurse Radiologist Lab. Technician		Vitamin K Antibiotics I.V. Glucose/ Fluids I.V. canula I.v. set	Radiant Warmer Saturation monitor Basic Lab. USG
2	Pediatrician with liver intensivists Pediatric surgeon Pediatric anaesthetists Radiologists Gastroenterologists Nuclear medicine		Higher antibiotics	NICU Pediatric O.T. Pathologist for biopsy Radiologists Nuclear medicine

Urinary tract infection and vesico ureteric reflux

ε. When to suspect and recognise

h) Introduction

Urinary tract infection (UTI) is a common condition affecting children and vesico ureteric reflux (VUR) is one common cause of recurrent UTI. UTI in the presence of infection results in kidney damage and one of the common causes of chronic renal failure in adult hood . UTI and VUR need prompt recognition to reduce morbidity and mortality in children.

(B) Case definition.

Vesico ureteric reflux may be primary due to short intramural course of ureters or it may be secondary to posterior urethral valves, Ureterocele. Neurogenic bladder

or it may be a part of duplex system of the kidney.

II . Incidence of the condition in our country

The exact incidence in our country is not known But from published data it is the risk of developing UTI before the age of 14 years is approximately 1% in boys and 3-5% in girls The incidence varies with age. During the first year of life, the male to female ratio is 3-5:1. Beyond 1-2 years, there is female preponderance with male to female ratio of 1:10. The incidence of VUR in children with UTI is approximately 30 to 50 % in the siblings of the index case shows an incidence of 20 to 30 %.

III . Differential diagnosis of VUR

- I.** Obstructive Mega ureters,
- II.** Ureterocele,
- III.** Urethral valves,

- IV. PUJ obstructions
- V. Ureteric stenosis,
- VI. Neurogenic bladder,
- VII. Posterior urethral valves with reflux
- VIII. Stones in the bladder and ureter.

IV. Prevention and counselling

VUR is congenital disease either primary or secondary to congenital outlet obstructions. As such prevention is not possible.

Presence of pelviureteric dilatation on antenatal scans needs evaluation soon after birth.

However UTI secondary to VUR can be prevented by prophylactic antibiotic therapy,

Periodical cultures and recognising infective episodes and treating them aggressively.

Mothers must be counselled on the need for long term chemo prophylaxis and have the urine culture done every febrile episode of the child with VUR. Early recognition can prevent damage and scarring to the kidney ,

As regards the parents can be counselled that Grade I to III is likely to disappear spontaneously in most cases within a period of two to three years. However spontaneous disappearance Gr IV and Gr V is likely to be less but can be given a chance of Chemo prophylaxis and observation for a period of two to three years.

Break through infections and fresh scars and structural abnormalities will be an indication for surgical intervention.

V. Optimal diagnostic criteria , investigations treatment and referral criteria.

Situation 1. At secondary hospital / Non metro situation ó optimal standards of treatment in situations where technology and resources are limited.

A. Clinical diagnosis

High degree of suspicion of VUR in all UTI patients.

Febrile episodes with anorexia , vomiting and shivering

Recurrent fever/ PUO

Recurrent vomiting

Failure to thrive.

Voiding dysfunction and dysuria

b. Investigations

Urine Routine and microscopy

Urine culture and sensitivity (MSSU)

Blood urea and serum creatinine

Ultra sonogram

Micturiting cysto urethrogram if expertise is available

C. Treatment:

If the urine culture is positive

c. Day care: No role for day-care

d. Out patient : UTI treated with appropriate oral antibiotics

e. Inpatient: UTI treated by appropriate antibiotics intravenously : Sick child

Based on culture and sensitivity report child needs admission . Appropriate antibiotics are chosen and administered for a period of 7 to 10 days intravenously.

It should be followed by oral chemo prophylaxis till the reflux subsides with periodical monitoring of the urine culture especially during febrile episodes.

In metro hospitals for the VUR no surgical intervention is done .

Ultrasonogram done shows some structural abnormalities , should be investigated further with Intravenous urogram and sent to higher centres for intervention.

Referral criteria

11. All cases of UTI who have not been evaluated with MCU should be referred to higher centre for further evaluation and plan of management.

12. All patients with VUR should be referred to higher centres for Radionuclide studies to see the differential renal function and assess the renal damage.

Lower grades of reflux with recurrent UTI and evidence of development of new scars and anatomical abnormalities need to be referred to higher centres for management.

(B) All cases of Gr IV and Gr V VUR should be referred to centres doing major paediatric surgical work as they do not undergo spontaneous resolution and require surgical or endoscopic management.

Situation 2

At super specialty at metro location where higher end technology is available

Investigations.

Routine urine examination and culture and sensitivity

Blood urea creatinine

Ultrasonogram

DMSA scan

IVU in selected cases to exclude upper urinary tract lesions

Bladder function Urodynamics studied in selected cases

Other investigations such as Plasma rennin activity and Genetic studies may be required in some cases.

Absolute indications for surgery

- i. Anatomic abnormalities of the bladder and VU junction
- j. Unresolving VU reflux
- k. Progressive renal injury
- l. Break through Pyelonephritis inspite of appropriate antibiotic prophylaxis
- m. Failure of renal and somatic growth

- n. Non compliance in medical management of the drug regime

Treatment

Primary treatment

Treatment of UTI with appropriate drug with appropriate dosage and period of time. Spontaneous resolution for most of the minor VUR and to a small extent Major VUR It happens over period of time which is usually 3 years.

Children need long term chemoprophylaxis & surveillance (BP monitoring), Somatic growth monitoring , renal function tests , urine analysis and Periodical cultures

Periodical assessment of renal condition with Ultra sonogram and Nuclear scans and MCU

Surgical treatment:

Type of surgery

Open surgery :- Reimplantation of ureters

Average stay 7 to 10 days

Has a success rate of 98 %

Child may need readmission for removal of stents if used for splinting the reimplanted ureters

Alternative management

There is a role for alternate procedures in select patients like

1.Circumcision

2.Endoscopic injection therapy

3 Diversion procedures like Ureterostomy and vesicostomy.

Long term management will include surveillance of the child and addressing bladder dysfunction if present.

Referrals

Even in Metro cities there are several levels of care. Surgical and endoscopic procedures should be done in institutions with proper cystoscopes for different age groups including

the neonates. C arm facilities and monitors to see the endoscopic procedures are necessary. Anaesthetist trained in paediatric anaesthesia is essential.

Who does what and timelines

- Doctor.
- Paediatric chief surgeon..... Main surgery
- Assistant surgeon... Assists the surgeon
- Scrub nurse
- Theatre technician
- X- Ray technician to monitor C Arm
- Ward staff

Further Reading and references

Consensus statement of management of Urinary tract infections

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Progress in Paediatric urology Edited by Minu bajpai. Volume 6

Vesico ureteric reflux

SITUATION	HUMAN RESOURCE	INVESTGATIONS	DRUGS AND CONSUMABLES	EQUIPMENT
1	Pediatrician, Nurse., ultrasonologist lab technician	Urinalysis urine culture ultrasound blood counts serum creatinine	Oral antibiotics	Ultrasound machine basic laboratory
2	Pediatric surgeon pediatric nephrologist/pediatricia n ultrasonologist, radiologist, nuclear medicine specialist ,	In addition to (1) fluoroscopy, nuclear medicine urodynamics	Oral antibiotics IV antibiotics material for IV access , material for radiological tests. Material for	In addition to (1) advanced laboratory, fluoroscopy facility nuclear medicine facility,

	urodynamics expert		nuclear medicine tests,	urodynamics operation theatre
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Neonatal Jaundice-Unconjugated hyperbilirubinemia

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NEONATAL JAUNDICE : UNCONJUGATED HYPERBILIRUBINEMIA

7. When to suspect / Recognize?

f. **Introduction:**

Neonatal jaundice is one of the most common neonatal problems. The most common cause of neonatal jaundice is physiologic jaundice. Although most newborns with jaundice are otherwise healthy, they need to be monitored because bilirubin is potentially toxic to central nervous system, causing bilirubin induced neuronal damage (BIND). Chronic BIND is also known as kernicterus. Kernicterus leads to permanent neurodevelopmental handicap. Physiologic jaundice is benign and self limiting, but pathologic jaundice can cause severe hyperbilirubinemia , which if not treated appropriately can result in kernicterus

g. **Case definition : For both situation of care (mentioned below)**

Neonatal jaundice : indicates presence of visible jaundice. Jaundice is visible in a neonate at bilirubin levels of 5 mg/dl

Neonatal hyperbilirubinemia : bilirubin > 15 mg/dl

Significant hyperbilirubinemia : jaundice requiring treatment (bilirubin > 17 mg/dl)

Severe hyperbilirubinemia : bilirubin > 20 mg / dl

Extreme hyperbilirubinemia : bilirubin > 25 mg / dl

Acute bilirubin encephalopathy : used to describe acute CNS manifestation of bilirubin toxicity.

Chronic bilirubin encephalopathy (Kernicterus): Chronic and permanent clinical sequelae of bilirubin toxicity

Bilirubin induced neurological dysfunction (BIND): changes associated with acute bilirubin encephalopathy

Physiological jaundice :

TERM infants :

- o. Onset on day 2-3 of life
- p. Reaches peak of 12mg/dl on day 4-5
- q. Subsides over 7-10 days

Preterm :

- φ. Onset on day 2 of life
- γ. Reaches peak of 15mg/dl on day 6-8
- η. Subsides over 10-14 days of life

There is no clear consensus on what is bilirubin cut off for physiological jaundice. However levels greater than 17 mg% are unlikely to be due to physiologic jaundice

Pathological jaundice :

- 8. Onset of jaundice before 24 hours of life
- 9. Rise in bilirubin of > 0.5 mg/dl/hour
- 10. Any level of bilirubin that requires phototherapy
- 11. Signs of underlying illness in an infant with jaundice (vomiting, lethargy, poor feeding, excessive weight loss, apnoea, tachypnoea, temperature instability)
- 12. Jaundice persisting more than 14 days in a preterm and term infant
- 13. Direct bilirubin of more than 2mg/dl or more than 15 % of total serum bilirubin.

8. INCIDENCE OF THE CONDITION IN OUR COUNTRY :

65 -70 % of all neonates have clinical jaundice . Incidence is higher among preterm neonates as compared to term neonates. Incidence of neonatal hyper bilirubinemia is 4.3- 6.5 % of all live births (NNPD 2005)

9. Differential diagnosis :

1. The most important aspect is to differentiate neonates with self limiting physiologic jaundice from those having neonatal jaundice due to underlying problems (pathologic jaundice), as the latter group can develop severe hyperbilirubinemia which can result in neuronal damage
2. Neonatal unconjugated hyperbilirubinemia should always be differentiated from neonatal cholestasis. One should suspect neonatal cholestasis if any neonate with jaundice has pale coloured stools and high coloured urine

Causes of indirect hyperbilirubinemia

- Fetomaternal blood group incompatibility ó ABO / Rh
- Hemolytic jaundice
- Drug induced ó Vitamin K, antimalarials, Oxytocin
- Collection of blood in the extravascular space (cephalhematoma)
- Polycythemia
- Metabolic and endocrine conditions
 - Galactosemia, Crigler Najjar syndrome, Hypothyroidism, hypopituitarism, Tyrosinosis
- Prematurity
- Infant of diabetic mother
- Sepsis

14. PREVENTION & COUNSELLING :

Primary prevention :

1. Screening for isoimmunisation:

All pregnant women should be tested for ABO/ Rh (D) typing

All neonates born to O blood group and Rh negative mothers should have a cord blood sample taken for blood grouping, typing and Coombs test (DCT)

2. Preventing hyperbilirubinemia :

Having hospital policies for detection for jaundice in newborn

- i) Protocols for assessment of jaundice at 12 hrly interval
- j) Identifying the babies at risk of developing hyperbilirubinemia
- k) Ensuring successful breast feeding

*****Babies at risk for developing hyperbilirubinemia are;**

Major:

- IX. Predischarge TSB (total serum bilirubin) in high risk zone
- X. Jaundice within first 24 hours
- XI. Blood group incompatibility with positive DCT
- XII. All preterm babies
- XIII. Previous sibling received photo therapy
- XIV. Cephalhematoma or significant bruising
- XV. Exclusive breast feeding , if nursing is not going on well and weight loss is excessive (greater >10%)

Minor:

- XVI. Predischarge TSB in the high intermediate risk zone
- XVII. Gestational age 37-38 weeks
- XVIII. Macrosomic infant of a diabetic mother

XIX. Maternal age >25 yrs

XX. Male gender

Counselling parents :

- a. It is essential to make sure that the parents are informed about newborn jaundice.
- b. Early discharge (<48 hours) is one of the reasons for missing neonates with hyperbilirubinemia (as breast feeding is not yet established and jaundice usually peaks at about 3 to 5 days)
- c. It is preferable to keep the mother and baby pair in the hospital at least for a period of 48 hours even for normal deliveries
- d. Those with major risk factors*** should definitely not be discharged early (preferably observed for 72 hours)
- e. If for any reason early discharge is planned, a pre-discharge bilirubin should be done and treatment planned as per the bilirubin nomogram (appendix 1) and frequent follow up is essential

15. Optimal diagnostic criteria: Investigations, treatment and referral criteria

**Situation 1: At secondary hospital / Non Metro situation:
Optimal standards of treatment in situation where technology
and resources are limited**

A) Clinical criteria:

Identify any risk factors in mother and baby

Clinical assessment using Kramer's criteria

Non invasive measurement of jaundice if available ó transcutaneous bilirubinometer

- **Lab measurement of bilirubin :**

Total serum bilirubin (TSB) and Direct bilirubin (daily)

More frequent bilirubin assessment (8-12 hrly) if hemolytic jaundice is suspected

Blood grouping/typing (ABO/Rh) of all mothers and neonates born to mothers with Rh negative and O positive mothers

Coombs test if mother is Rh negative or O positive

Complete hemogram including Peripheral smear , Reticulocyte count if hemolytic jaundice is suspected or neonate requires phototherapy

Jaundice present beyond 2 weeks or sick infants need further evaluation

C) Treatment:

No role of outpatient or day care treatment

If a neonate is referred from outside, neonate needs to be assessed for level of jaundice and assessment of risk factors needs to be done***

The following will need to be admitted and bilirubin estimation should be done

- Any term neonate having clinical jaundice till lower abdomen and beyond
- Any preterm neonate having jaundice
- Any neonate having jaundice in the first 24 hrs
- Any neonate having jaundice beyond 2 weeks or has pale coloured stools

INPATIENT TREATMENT

1. AAP guide lines for management of hyperbilirubinemia in baby >_35 weeks (Appendix 2)

The bilirubin level is estimated and plotted on the chart based on age of the baby. Decision is taken depending on which part of the graph the value falls

- In addition to providing phototherapy for those who require the following policy needs to be practiced for all intramural neonates
- Promote & support successful breast feeding
- Establish nursery protocols for identification and evaluation of hyperbilirubinemia
- Assess all neonates for presence of risk factors
- Only visual estimation of degree of jaundice can lead to errors in darkly pigmented infants, therefore frequent bilirubin estimations will have to be done
- Perform systemic assessment on all infants prior to discharge for the risk of hyperbilirubinemia
- Do a serum bilirubin for infants discharged before 48 hours
- Interpret all babies bilirubin levels according to infant age, in hour specific nomogram
- Provide appropriate follow up based on the time of discharge and risk assessment.
- Treat newborn when indicated with phototherapy/ exchange transfusion

Phototherapy

Indications for phototherapy in neonates above 35 weeks is based on the chart provided (appendix 2)

Indications in preterm babies

In preterm neonates < 35 weeks there are no charts

Guidelines for phototherapy in thus group is as follows

< 1000gms ? prophylactic phototherapy

1000 to 1500 gms 7 to 9 mg%

1500 ó 2000gms 10 to 12 mg%

2000 ó 2500gms 13 ó 15 mg%

Duration of phototherapy : to be given till the bilirubin value reaches safe level as per chart. It is advisable to keep the neonate in hospital for a period of 12 to 24 hrs or repeat a bilirubin value 12 to 24 hrs after stopping phototherapy as rebound hyperbilirubinemia can occur, particularly in neonates with hemolytic jaundice

Types of light: Single or double surface phototherapy .

A combination of special blue (TL20) lights and white lights

Do not use white lights covered with blue paper as they are ineffective

IT IS IMPORTANT TO CHANGE LIGHTS AFTER EVERY 1000 HOURS OF USE

EXCHANGE TRANSFUSION CAN BE DONE AT A SECONDARY LEVEL ONLY IF EXPERTISE IS AVAILABLE

If expertise is not available early referral of high risk neonates is recommended

- **Referral criteria:** Early referral
 - Presence of major risk factors (Rh/ABO)

- Onset of jaundice within 24 hours
- Rapid rise of bilirubin in spite of phototherapy
- TSB is nearing Exchange transfusion range
- Associated other morbidity

Situation 2

Tertiary centre

a) Clinical diagnosis :

Same as Explained

c) Investigation :

Non invasive mode of detection of jaundic

Transcutaneous bilirubinometry

These instruments gives fairly accurate estimates of TSB in term/near term, values within 2-3mg/dl

They have been considered as a pre-discharge screening tool to identify at risk infants

Lab investigation :

As above

Additional investigations

- G6PD
- T3 T4 TSH
- Bilirubin monitoring 6-8 hourly in neonates with hemolytic jaundice or 12 to 24 hrly in other neonates

Treatment :

Phototherapy : started when the bilirubin value crosses the percentile charts. Appendix 2

The lights which can be used for the treatment are:

Special blue light (TL 20) -

(acceptable range for flux should be 8-10 microwatts/cm² /nm and for intensive photo therapy ; >30 microwatts/cm² /nm

Fiberoptic (bili blanket)

LED (light emitting diode)

Exchange transfusion

Is indicated if bilirubin rises beyond recommended levels as per chart Appendix 3

It is indicated at earlier levels in preterm neonates and neonates with immune hemolysis

Guideline for preterms

< 1000gms	10 to 12 mg %
1000 to 1500 gms	12 to 15 mg%
1500 ó 2000gms	15 to 18 mg%
2000 ó 2500gms	18 to 20 mg%

Rh or ABO immune hemolysis

Bilirubin values of 10 mg% within 24 hrs of age

15 mg% between 24 to 48 hours of age

In all other situations - as per chart (appendix 3)

Other pharmacological treatment ;

- Intravenous immunoglobulin(IVIG) is useful as it inhibits hemolysis, may decrease the need for ET but does not eliminate it completely. IVIG can be used in neonates either Rh or ABO isoimmunization (dose)
- **Tin mesoporphyrin** is at 6 micro mol/kg more effective than special blue light . Human studies have shown that a single IM dose of 6 microM/kg eliminates the need of phototherapy in the postnatal period. (Not yet available)

2) Standard operational procedure

Inpatient :

- **Phototherapy**
- Exchange transfusion : whole blood cross matched against mothers group ,matched to baby blood group .volume decided by the weight of the baby. It is an invasive procedure requires cannulation of the umbilical vein/any other peripheral vein.

Out patient : Sampling blood for investigations,cross matching , no procedure on outpatient basis

Referral criteria :

No referral from tertiary centre

13. WHO DOES WHAT ? and TIMELINES

Doctor :

Identification of the babies with risk factors

To follow up the cord blood group as early as possible after birth,initiate treatment early if required especially if there is O/ Rh negative history

Daily assessment for jaundice clinically if required investigation

Decision regarding Photo therapy / exchange transfusion

Perform Exchange transfusion

Nurse :

To assess clinically for jaundice when checking 6 -8 hrly vitals

To inform doctors and to do trans cutaneous bili check or send blood for bilirubin

Assessment of clinical status , hydration, side effects while the baby on the photo therapy.

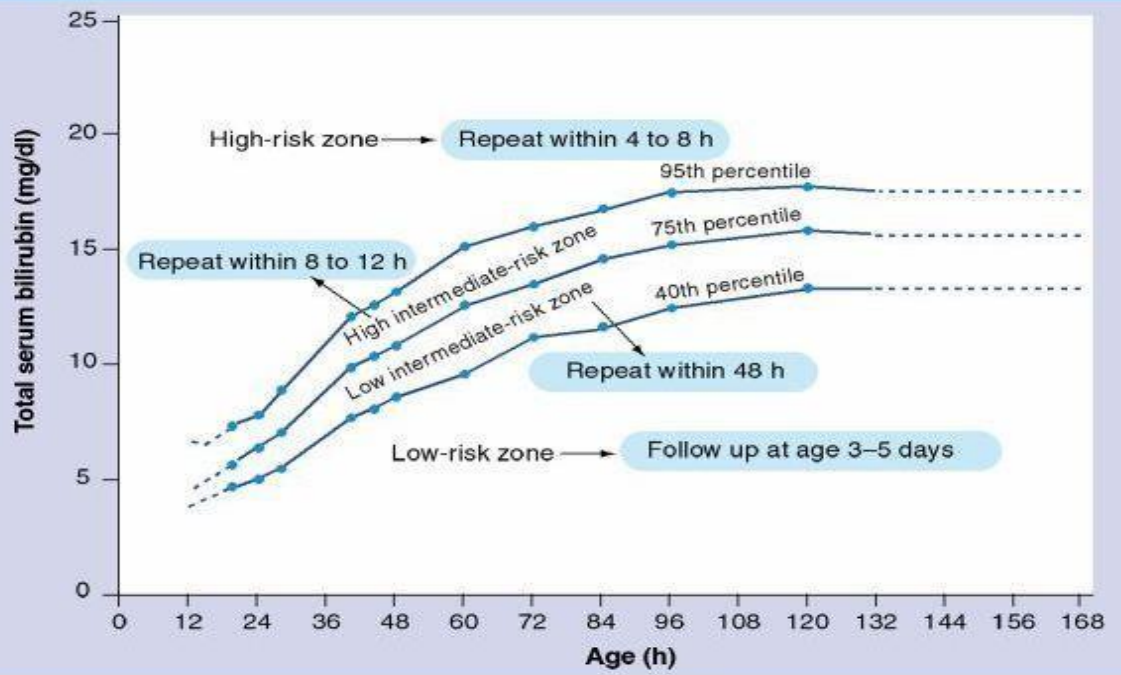
Technician :

To find the bilirubin value as soon receiving the sample in a risk baby and immediate information if abnormal

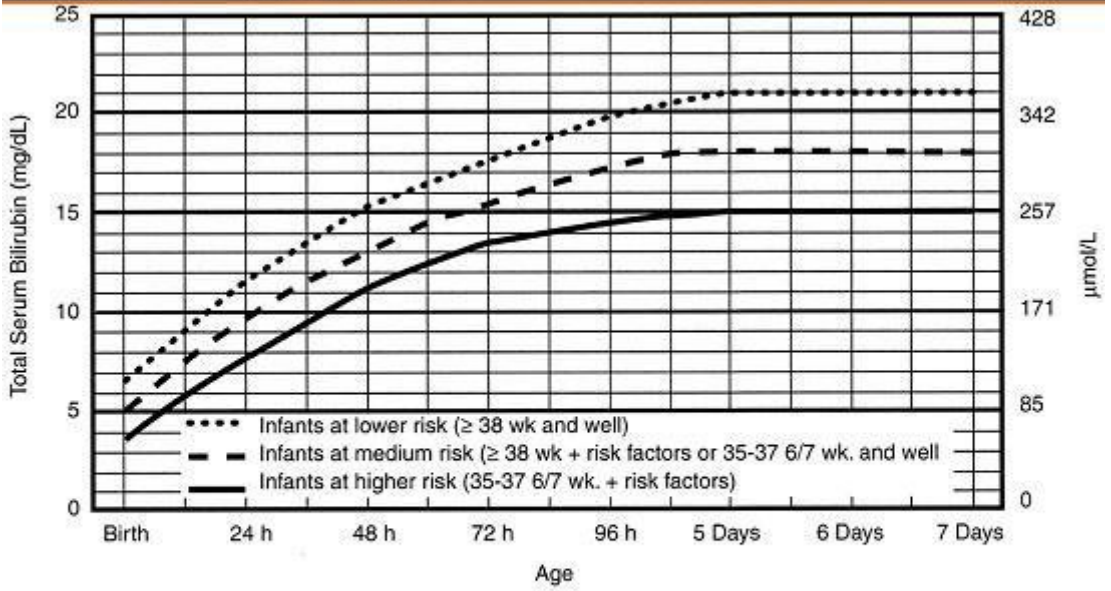
Information on the abnormal blood group ,DCT report to concerned people

7) Further reading/ references:

6. Guha's Neonatology principles and practice 3rd edition 2005
7. Neonatal pathology and management of the newborn by Avery , 6th edition
8. Neonatal perinatal medicine- Avroy A Fanaroff
9. Fanaroff and Martin's Neonatal- Perinatal medicine- Diseases of the fetus and infant- 8th edition

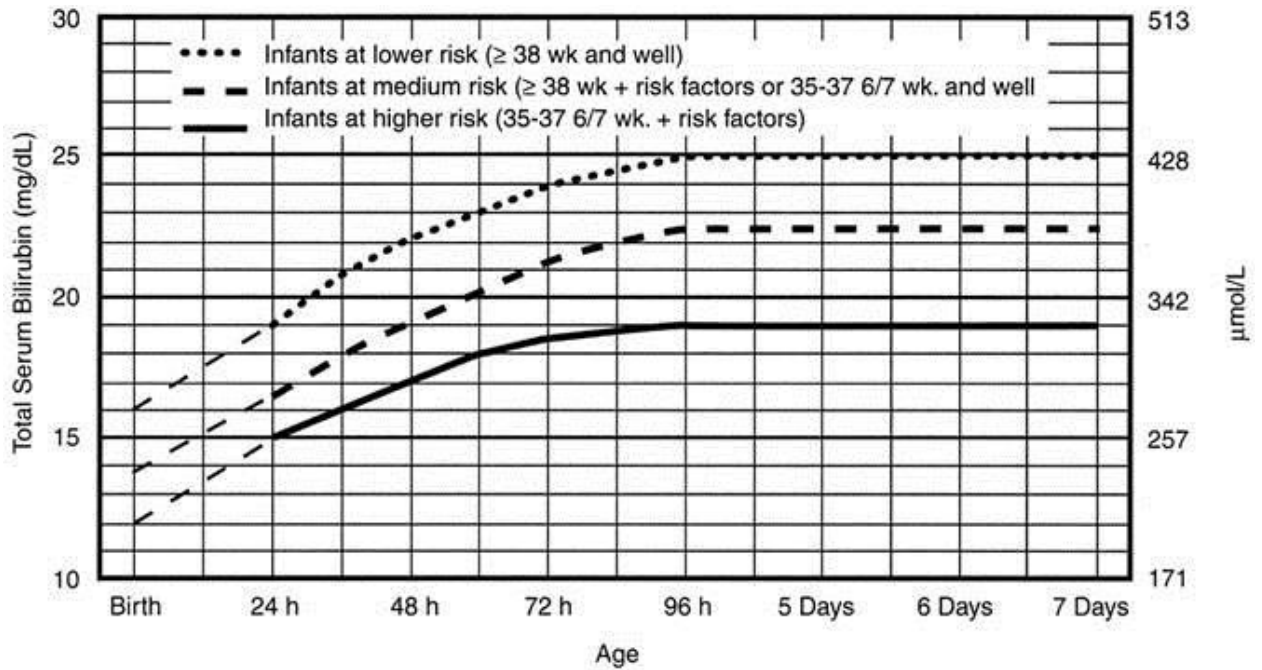


Appendix 1



- Use total bilirubin. Do not subtract direct reacting or conjugated bilirubin.
- Risk factors = isoimmune hemolytic disease, G6PD deficiency, asphyxia, significant lethargy, temperature instability, sepsis, acidosis, or albumin $< 3.0\text{g/dL}$ (if measured)
- For well infants 35-37 6/7 wk can adjust TSB levels for intervention around the medium risk line. It is an option to intervene at lower TSB levels for infants closer to 35 wks and at higher TSB levels for those closer to 37 6/7 wk.
- It is an option to provide conventional phototherapy in hospital or at home at TSB levels 2-3 mg/dL (35-50mmol/L) below those shown but home phototherapy should not be used in any infant with risk factors.

Appendix 2



- The dashed lines for the first 24 hours indicate uncertainty due to a wide range of clinical circumstances and a range of responses to phototherapy.
- Immediate exchange transfusion is recommended if infant shows signs of acute bilirubin encephalopathy (hypertonia, arching, retrocollis, opisthotonos, fever, high pitched cry) or if TSB is ≥ 5 mg/dL ($85 \mu\text{mol/L}$) above these lines.
- Risk factors - isoimmune hemolytic disease, G6PD deficiency, asphyxia, significant lethargy, temperature instability, sepsis, acidosis.
- Measure serum albumin and calculate B/A ratio (See legend)
- Use total bilirubin. Do not subtract direct reacting or conjugated bilirubin
- If infant is well and 35-37 6/7 wk (median risk) can individualize TSB levels for exchange based on actual gestational age.

Appendix 3

Methods of giving Phototherapy

Baby fulfills the criteria for phototherapy

- To make sure that the lights are in usable condition (been used not more than 1000 hrs or > 3 months whichever is earlier)
- If double light phototherapy is given , infant lies on a fiberoptic blanket with conventional phototherapy overhead or a double surface phototherapy can be given
- To place the light as close to the baby as possible, if double light phototherapy not given, position of the baby to be changed frequently
- If an incubator is used there should be a 5-8 cm space between it and the lamp cover to prevent over heating
- The most effective lights for phototherapy are those with output 425-475 nm
- The baby should be naked with eyes and genitalia covered
- Temperature should be monitored and maintained
- Infant should be weighed daily
- To monitor the hydration status of the baby and adjust the fluid/ feeds accordingly
- Frequent breast feeding, additional oral fluids only if significant weight loss is present (> 10% wt loss)
- Skin colour is not a guide to hyperbilirubinemia in infants on phototherapy ,hence bilirubin should be monitored every 12 hrs
- Once satisfactory decline in bilirubin has occurred, infants can be removed from phototherapy for feeding

Exchange transfusion

- To make sure the baby fulfills the criteria for exchange transfusion
- **Choice of blood:**
Fresh, less than 7 days old, irradiated (if possible), whole blood
In Rh hemolytic disease, blood is prepared before delivery, type O Rh negative crossed matched against mother. If the blood is obtained after delivery to be crossed against baby OR ABO group of baby and Rh negative blood
ABO incompatibility: O Rh negative or Rh compatible with the baby and mother, crossed matched against mother and infant and have low

- titer of anti A or anti B antibodies Usually type O cells are used with AB plasma to ensure that no anti A or anti B antibodies
- **Volume for Exchange transfusion** involves double volume of the infants blood that is $80 \text{ ml/ kg} \times 2 = 160 \text{ ml/kg}$ (in preterm neonates blood volume = 100 ml/kg)
 - **Technique :**

Procedure done under servocontrolled warmer and cardiac and blood pressure monitoring,

- Equipment and personnel for resuscitation should be readily available
- an assistant to record volumes of blood, observe and check the vitals
- Infants arm and legs are properly restrained
- Blood warmed to 37 degree centigrade
- Sterile technique used during the whole procedure, Umbilical vein cannulated , position of the catheter to be checked by X óray
- Procedure done using 3 way valve assembly, most often push pull technique used
- The total time taken for the procedure to be not more than 1-1 ½ hrs,
- Photo therapy to be continued after the procedure
- CVP should be measured before and after the procedure
- During the procedure care to be taken to monitor the baby for vitals
- The amount of blood per aliquot depends on weight of the baby which is as follows

<1000 gms - 3ml
1000- 1500 gms - 5 ml
1500-2000 gms -10 ml
2000-3500 gms - 15 ml
>3500 gms -20 ml

Standard Treatment Guidelines Group

Pediatrics and Pediatric Surgery

Date: 28.02.2011

NEONATAL INTESTINAL OBSTRUCTION

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XXI. WHEN TO SUSPECT/ RECOGNIZE?

a. Introduction:

Neonatal Intestinal Obstruction (NIO) is a common condition affecting neonates and requires prompt recognition and appropriate specialist treatment to save these babies and to reduce avoidable morbidity & mortality

b. Case definition:

Intestinal Obstruction in a new born child caused by intrinsic or extrinsic factors involving the stomach / duodenum / jejunum / ileum / any part of the colon.

XXII. INCIDENCE OF THE CONDITION IN OUR COUNTRY

- 1. The Exact incidence of the condition in our population is difficult to ascertain. However it may be approximately assessed to be 14 per 1,000 live births**

DIFFERENTIAL DIAGNOSIS

- a. Gastric Outlet Obstruction - Pyloric / Antral web / Hypertrophic Pyloric Stenosis**
- b. Malrotation**
- c. Duodenal Atresia**
- d. Jejuno-ileal Atresia**
- e. Meconium Ileus**
- f. Colonic Atresia**
- g. Hirschprung's Disease**
- h. Meconium Plug Syndrome**
- i. Necrotising Enterocolitis (NEC)**

- j. **Medical conditions mimicking Neonatal intestinal obstruction like Neonatal Sepsis with ileus, Hypothyroidism etc.**
- k. **Other rare conditions like intestinal duplications, intra-abdominal cysts, congenital bands etc.**

PREVENTION AND COUNSELING

- **As this is often a congenital disease, prevention is impossible.**
- **However, the obstetricians at all levels need to have a high index of suspicion about the possibility of a GI obstruction when polyhydramnios is detected on antenatal scans**
- **When a fetus is suspected to have NIO, the counseling can be done to have a discussion with the obstetrician, the pediatrician & the pediatric surgeon and to plan the delivery to be done at a place where neonatal surgery is safely feasible**

16. OPTIMAL DIAGNOSTIC CRITERIA, INVESTIGATIONS, TREATMENT & REFERRAL CRITERIA

***Situation 1: At Secondary Hospital/ Non-Metro situation: Optimal Standards of Treatment in Situations where technology and resources are limited**

- **Clinical Diagnosis:** Any child with any of the following criteria should be suspected to have NIO
 10. Bilious Vomiting (green colour)
 11. Abdominal distention
 12. Visible peristalsis
 13. Not passed meconium beyond 48 hours after birth (72 hours in preterm babies)
 14. Passing abnormal stool (pellets / mucus / mucus plug)
 15. Feed intolerance
 16. Upper or Lower Gastrointestinal Bleeding.

Persistent Non-bilious vomiting

Investigations:

- a. Plain X-Ray Abdomen (preferably erect)
- b. Air contrast X-Ray - A naso-gastric tube is inserted prior to shifting the baby for the X-Ray. About 20 cc of Room air is insufflated through the naso-gastric tube and the X-Ray is taken. (This is to highlight the stomach, duodenum and the proximal jejunum and to rule out obstruction at this level.). Make sure the insufflated air is aspirated out immediately after the X-Ray is taken and the NG tube is left for drainage
- c. Ultrasonography, if indicated.

▪ **Treatment:**

Standard Operating procedure

10. Day Care - No role for treatment on day care basis
11. Out Patient - No role for treatment on out-patient basis
12. In Patient -
 - **Gastric Deompression**
 - Place a No. 8 infant feeding tube through the nasogastric route. In very small babies, oro-gastric tube can be utilised if the nostrils are small. The baby should be kept nil orally.
 - Ensure that the tube is correctly placed in the stomach and that it is patent.
 - Gently aspirate the tube hourly and connect to continuous drainage.
 - Maintain accurate chart to monitor the colour and volume of the aspirates
 - i. **Temperature Maintenance**
 - Keep the child warm using an incubator / warmer and keeping the room ambient temperature high. A room warmer can be used in winter seasons
 - **Intravenous Fluids to maintain Hydration / glucose levels.**
 - The choice of IV fluids in the first 48 hours is 10% Dextrose @ 80 ml/ kg / day.

- After 48 hours of birth, the IV fluid of choice would be Isolyte P to be run @ 100 - 120 ml / kg / day (the rate will vary as per the gestation of the baby and other factors as determined by the pediatrician).
- The fluid should be infused using 'Burette' / 'Pediatric Drip Chamber set' in order to avoid over or under infusion. Microdrip sets with 'dosiflow' regulators may be used if these are not available.
- **I.V. Antibiotics. Broad spectrum antibiotics to cover gram positive, gram negative and anerobic bacteria should be used. Some of the suggested combinations are:**
 1. Ampicilin / Gentamicin / Metronidazole
 2. Cefotaxime / Amikacin / Metronidazole
 3. Co-Amoxiclav / Amikacin / Metronidazole
 4. The choice of the antibiotic will vary on a lot of factors and can be suitably chosen by the treating clinician .
- ii. **Preferable Investigations**
 1. Serum glucose / Electrolytes
 2. BUN / Serum Creatinine
 3. Hemoglobin & Blood Counts
 4. Blood culture
 5. Ultrasonography of the Abdomen

- 6.
- 7.
- 8.

b) Referral criteria:

- a. Ideally, all cases of neonatal intestinal obstruction should be transferred to hospitals with level 2 / 3 Neonatal intensive care facilities.
- b. However, in situations where a qualified pediatric surgeon is available, these children can be handled provided the hospital has the following facilities
 - i. Neonatal nursery with the availability of full-time trained neonatal / pediatric nurses
 - ii. Round the clock availability of Pediatrician
 - iii. Warming system for the baby - Radiant warmer/ Incubator etc/
 - iv. Operation Theatre well equipped with Monitors for ECG / Pulse Oximetry / Baby warming systems and other facilities for operating on a small baby
 - v. A Well Trained Anesthesiologist with adequate exposure to neonatal anesthesia
 - vi. Facilities for post-operative monitoring of the baby - warmer, multi-system monitor, resuscitation equipment etc.
- c. Mode of transportation & Precautions during transfer
 - i. Keep the child warm using clean blankets/ thermocol boxes, cotton padding etc, Keep the NGT open & connect to continuous drainage. Strictly Avoid oral / NGT feeds
 - ii. Maintain the patency of IV line by flushing it before transportation and run fluids at the pre-determined rate if the travel is expected to last more than a few hours
 - iii. It is preferable to have a trained paramedical / medical supervision during transportation . The person should preferably be trained in basic neonatal care / neonatal resuscitation methods, handling medical equipment like those required for airway maintenance / suction etc.
 - iv.

***Situation 2: At Super Specialty Facility in Metro location where higher-end technology is available**

a) **Clinical Diagnosis:** Same as earlier

b) **Investigations:**

a. **Blood:**

- i. CBC / CRP / Blood Culture
- ii. S. Electrolytes / BUN / Creatinine
- iii. Arterial Blood Gas
- iv. Blood Grouping & Rh typing /
- v. Serum Bilirubin
- vi. Other blood tests as deemed necessary by the neonatologist.

b. **Imaging: (Depending on the working diagnosis)**

- i. Plain X-Ray of Abdomen in all cases
- ii. Air contrast X-Ray - if upper GI obstruction is suspected.
- iii. Upper G.I. barium study - if upper GI obstruction is suspected.
- iv. Contrast (Gastrograffin preferably) Enema - if lower GI obstruction is suspected.
- v. Ultrasonography of Abdomen / Pelvis
- vi. CT scanning / MRI - in rare instances

c. **Additional** Screening for Associated anomalies may be required in select cases

- i. 2-D Echocardiography
- ii. Renal Ultrasonography
- iii. Chromosomal & Metabolic screening
- iv. Any other as indicated

c) **Treatment:**

Standard Operating procedure

- a. Out Patient - no role**
- b. Day Care - No role**
- c. In Patient:**
 - i. Initial resuscitation / Stabilisation**
 - ii. IV Fluids / Antibiotics as indicated earlier**
 - iii. Pre-operative Preparation - This will by and large depend on the condition of the baby and if any pre-existing morbidity is present and will be handled by the neonatologist.**
 - iv. Operative plan - This will depend on the diagnosis made about the level of obstruction. The possibilities include**
 - 1. Simple laparotomy + Ladd's procedure / release of bands / Pyloromyotomy etc.**
 - 2. Laparotomy + Resection anastomosis**
 - 3. Laparotomy + Resection + Ileostomy / colostomy**
 - 4. Laparotomy + pull-thorough**
 - 5. One stage pull-through for Hirschprung's Disease**
 - 6. Laparotomy + Other procedures as per variations in the operative findings.**
 - i. Post-operative Care - This will again have to be tailored to suit the child's condition and requirements. The probable supportive measures will include:**
 - 1. IV Fluids with possible transfusion of blood products like Packed Cells / Plasma / etc.**
 - 2. IV Antibiotics- as suggested earlier and as dictated by various factors influencing the decision making process.**
 - 3. Inotropic Support - Dopamine / Dobutamine / Noradrenaline**
 - 4. Ventilatory Support**
 - 5. Parenteral Nutrition**
 - 6. Advanced Support like peritoneal dialysis**
 - 7. Vascular access**
 - ii. Some children may also require Additional Surgery like:**
 - 1. Re-look laparotomy**
 - 2. Definitive surgery**

- 3. Explorations for complications like anastomotic leak, adhesive obstruction / dehiscence
- 4. Ilesotomy / colostomy closure
- iii. Other events during hospitalisation complicating the clinical course during hospitalisation = like renal failure, nutritional support, colostomy / ileostomy care etc.
- iv. Maternal support during hospitalisation including rooming in while feeding is initiated

d) **Referral criteria:**

- a. Even within the metro cities, as there are several levels of hospital care available, we recommend that new born babies with surgical problem should be handled by only those hospitals with reasonably good neonatal care (level 2 & 3) facilities with the availability of qualified Pediatric Surgeon and an experienced pediatric anesthetist.
- b. However, depending on the nature of the disease and the general condition of the baby, decision may be taken to handle the baby in centres with less than optimal facilities in Metro cities if there is a genuinely good cause to believe that good surgical and post-operative care can be extended to the child without much detriment to the baby
- c. In any situation, **after the initial resuscitation**, if the general condition of the baby is poor or if there is a possible necessity of ventilatory support or specialised treatment, it will be necessary to shift the baby to a higher centre where such facilities are available, ensuring safe transportation of the baby

17. WHO DOES WHAT? and TIMELINES

a. **Doctor**

i. **Pediatrician:**

- 1. Initial assessment and day to day care of the baby,
- 2. Early involvement of a pediatric surgeon and regular co-ordination with him/ her
- 3. taking appropriate decisions & involving the various specialists as indicated

ii. **Resident / Registrar**

1. Periodic assessment of the patient and regular reporting to the specialists
 2. Carry out the orders of the Pediatrician / Pediatric Surgeon in charge of the patient
 3. To ensure that all the orders are properly carried out by the nursing and other paramedical personnel
 4. Blood sampling and vascular access
- iii. Pediatric Surgeon
1. Prompt assessment of the baby on referral and to formulate an appropriate plan of action
 2. Co-ordinating with the anesthetist and the other Operation Theatre personnel for the proposed surgery
 3. Performing the appropriate surgery and to make reasonable efforts for a smooth post-operative recovery.
 4. Post-operative care & Daily assessment with regard to the post-operative recovery
 5. Vascular access
 6. Take decisions with regard to the daily progress and further interventions as and when indicated
- iv. Anesthetist
1. Suitable pre-operative preparation
 2. Appropriate anesthetic care and smooth post-operative recovery
 3. Co-ordination with the other clinicians involved in the care of the child
- v. Neonatologist
1. Initial assessment and day to day care of the baby,
 2. Early involvement of a pediatric surgeon and regular co-ordination with him/ her
 3. Taking appropriate decisions & involving the various specialists as indicated
 4. Other specialist interventions AS AND WHEN NEEDED like :
 - a. Vascular access
 - b. Umbilical Venous / Arterial cannulation
 - c. Peritoneal dialysis
 - d. Enteral / Parenteral nutrition

- a. Nurse**
 - i. Nursing care of the baby**
 - ii. Following all the instructions of the attending doctors**
 - iii. Close co-ordination with all the departments**
 - iv. Maintaining the records of the children upto date**
- a. Emergency Room**
 - i. Ward**
 - ii. Neonatal Intensive Care Unit**
 - iii. Operation Theatre**
 - iv. Post-operative Recovery**
- a. Technician**

PROTOCOL FOR NEONATAL SEIZURES

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Neonatal Seizures

(Seizure protocol adapted from: Sankar JM, Agarwal R, Deorari A, Paul VK. AIIMS Protocol on management of neonatal seizures. Available at: http://www.newbornwhocc.org/pdf/Seizures_2010_270810.pdf)

a) **WHEN TO SUSPECT/ RECOGNIZE?**

a) **Introduction:**

Neonatal seizures (NS) are the most frequent and distinctive clinical manifestation of neurological dysfunction in the newborn infant. Infants with NS are at high risk of neonatal death or neurological impairment and epilepsy disorders in later life. Though, mortality due to NS has decreased over the years from 40% to about 20%, the prevalence of long-term neurodevelopment sequelae has largely remained unchanged at around 30%.² Improper and inadequate management of seizures could be one of the major reasons behind this phenomenon.

b) **Case definition and classification:**

A seizure is defined clinically as a paroxysmal alteration in neurologic function, i.e. motor, behavior and/or autonomic function.

Four types of NS have been identified- subtle, clonic, tonic and myoclonic. Myoclonic seizures carry the worst prognosis in terms of neurodevelopmental outcome and seizure recurrence. Focal clonic seizures have the best prognosis.

In secondary level hospital	At tertiary care centers
A clinical definition should be used for recognition and classification of NS	The diagnosis essentially is clinical. However sophisticated investigations such as EEG may be employed for confirmation and further classification.

b) INCIDENCE OF THE CONDITION IN OUR COUNTRY

The incidence of NS is 2.8 per 1000 in infants with birth weights of more than 2500 g; it is higher in preterm low birth weight neonates ó as high as 57.5 per 1000 in very low birth weight infants.

c) DIFFERENTIAL DIAGNOSIS

The most common causes of seizures as per the recently published studies from the country are hypoxic ischemic encephalopathy, metabolic disturbances (hypoglycemia and hypocalcemia), and meningitis.

HIE secondary to perinatal asphyxia is the commonest cause of NS. Most seizures due to HIE (about 50-65%) start within the first 12 hrs of life while the rest manifest by 24-48 hours of age. Common metabolic causes of seizures include hypoglycemia, hypocalcemia, and hypomagnesemia. Meningitis should be excluded in all neonates with seizures. Meningoencephalitis secondary to intrauterine infections (TORCH group, syphilis) may also present as seizures in the neonatal period. Seizures due to subarachnoid, intraparenchymal or subdural hemorrhage occur more often in term neonates, while seizures secondary to intraventricular hemorrhage (IVH) occur in preterm infants. Cerebral dysgenesis and neuronal migration disorders are rare

causes of seizures in the neonatal period.

a) PREVENTION AND COUNSELING

Good obstetric and neonatal care would go a long way in prevention of neonatal seizures. Screening and management of polycythemia and hypoglycemia can prevent seizure occurrence due to these reasons. Avoiding animal milk feeding by exclusive breastfeeding may reduce seizures due to late onset hypocalcemia.

b) OPTIMAL DIAGNOSTIC CRITERIA, INVESTIGATIONS, TREATMENT & REFERRAL CRITERIA

Approach and diagnosis

In secondary level hospital	At tertiary care centers
<ul style="list-style-type: none">• Clinical evaluation• Measurement of blood glucose, calcium, hematocrit• Lumbar puncture	<ul style="list-style-type: none">• Clinical evaluation• Measurement of blood glucose, calcium, hematocrit• Lumbar puncture• Neuroimaging (USG brain for preterm; CT/MRI for term baby)• EEG

Detailed approach to an infant with neonatal seizures^{1, 4-6}

1. History

Seizure history: A complete description of the seizure should be obtained from the parents/attendant. History of associated eye movements, restraint of episode by passive flexion of the affected limb, change in color of skin (mottling or cyanosis), autonomic phenomena, and whether the infant was conscious or sleeping at the time of seizure should be elicited. The day of life on which the seizures occurred may provide an important clue to its diagnosis. While seizures occurring on day 0-3 might be related to

perinatal asphyxia, intracranial hemorrhage, and metabolic causes, those occurring on day 4-7 may be due to sepsis, meningitis, metabolic causes, and developmental defects.

Antenatal history: History suggestive of intrauterine infection, maternal diabetes, and narcotic addiction should be elicited in the antenatal history. A history of sudden increase in fetal movements may be suggestive of intrauterine convulsions.

Perinatal history: Perinatal asphyxia is the commonest cause of neonatal seizures and a detailed history including history of fetal distress, decreased fetal movements, instrumental delivery, need for resuscitation in the labor room, Apgar scores, and abnormal cord pH (<7) and base deficit (>10 mEq/L) should be obtained. Use of a pudendal block for mid-cavity forceps may be associated with accidental injection of the local anesthetic into the fetal scalp.

Family history: History of consanguinity in parents, family history of seizures or mental retardation and early fetal/neonatal deaths would be suggestive of inborn errors of metabolism. History of seizures in either parent or sib(s) in the neonatal period may suggest benign familial neonatal convulsions (BFNC).

2. Examination

Vital signs: Heart rate, respiration, blood pressure, capillary refill time and temperature should be recorded in all infants.

General examination: Gestation, birth-weight, and weight for age should be recorded as they may provide important clues to the etiology. For example, seizures in a term well baby may be due to subarachnoid hemorrhage while seizures in a large for date baby may be secondary to hypoglycemia. The neonate should also be examined for the presence of any obvious malformations or dysmorphic features.

CNS examination: Presence of a bulging anterior fontanel may be suggestive of

meningitis or intracranial hemorrhage. A detailed neurological examination should include assessment of consciousness (alert/drowsy/comatose), tone (hypotonia or hypertonia), and fundus examination for chorioretinitis.

Systemic examination: Presence of hepatosplenomegaly or an abnormal urine odor may be suggestive of IEM. The skin should be examined for the presence of any neurocutaneous markers. Presence of hypopigmented macules or ash-leaf spot would be suggestive of tuberous sclerosis.

3. Investigations

In secondary level hospitals	At tertiary care centers
<ul style="list-style-type: none"> • blood sugar measurement • sepsis work up and LP • rule out polycythemia 	<ul style="list-style-type: none"> • blood sugar, calcium and electrolyte measurement • sepsis work up and LP • rule out polycythemia • neuroimaging • metabolic work up • EEG

Essential investigations: Investigations that should be considered in all neonates with seizures include blood sugar, serum electrolytes (Na, Ca, Mg), cerebrospinal fluid (CSF) examination, cranial ultrasound (US), and electroencephalography (EEG). CSF examination should be done in all cases as seizures may be the first sign of meningitis. It should not be omitted even if another etiology such as hypoglycemia is present because meningitis can often coexist. CSF study may be withheld temporarily if severe cardiorespiratory compromise is present or even omitted in infants with severe birth asphyxia (documented abnormal cord pH/base excess and onset within 12-24 hrs). An arterial blood gas (ABG) may have to be performed if IEM is strongly suspected.

One should carry out all these investigations even if one or more investigations are positive, as multiple etiologies may coexist, e.g. sepsis, meningitis and hypoglycemia.

Imaging: Neurosonography is an excellent tool for detection of intraventricular and parenchymal hemorrhage but is unable to detect SAH and subdural hemorrhage. It should be done in all infants with seizures. CT scan should be done in all infants where an etiology is not available after the first line of investigations. It can be diagnostic in subarachnoid hemorrhage and developmental malformations. Magnetic resonance imaging (MRI) is indicated only if investigations do not reveal any etiology and seizures are resistant to usual anti-epileptic therapy. It can be diagnostic in cerebral dysgenesis, lissencephaly, and other neuronal migration disorders.

Electroencephalogram (EEG): EEG has both diagnostic and prognostic role in seizures. It should be done in all neonates who need anticonvulsant therapy. Ictal EEG may be useful for the diagnosis of suspected seizures and also for diagnosis of seizures in muscle-relaxed infants. It should be done as soon as the neonate is stable enough to be transported for EEG, preferably within first week. EEG should be performed for at least one hour.⁹ Inter-ictal EEG is useful for long-term prognosis of neonates with seizures. A background abnormality in both term and preterm neonates indicates a high risk for neurological sequelae. These changes include burst-suppression pattern, low voltage invariant pattern and electro-cerebral inactivity.

Management

In secondary level hospitals	At tertiary care centers
<ul style="list-style-type: none"> • Stabilize the baby • Correct hypoglycemia or hypocalcemia, if present • Anti-epileptic drug therapy (AED): 	<ul style="list-style-type: none"> • Stabilize the baby • Correct hypoglycemia or hypocalcemia, if present • Anti-epileptic drug therapy (AED):

<p>phenobarbitone is the first choice</p> <ul style="list-style-type: none"> • Refer if intractable seizures, or baby requires ventilation 	<p>phenobarbitone is the first choice</p> <ul style="list-style-type: none"> • Use additional drugs to control seizures • Management of concurrent illnesses
---	--

Detailed approach to an infant with neonatal seizures^{1, 4-6}

1. Initial medical management:

The first step in successful management of seizures is to nurse the baby in thermoneutral environment and to ensure airway, breathing, and circulation (TABC). Oxygen should be started, IV access should be secured, and blood should be collected for glucose and other investigations. A brief relevant history should be obtained and quick clinical examination should be performed. All this should not require more than 2-5 minutes.

2. Correction of hypoglycemia and hypocalcemia:

If glucoStix shows hypoglycemia or if there is no facility to test blood sugar immediately, 2 ml/kg of 10% dextrose should be given as a bolus injection followed by a continuous infusion of 6-8 mg/kg/min.

If hypoglycemia has been treated or excluded as a cause of convulsions, the neonate should receive 2 ml/kg of 10% calcium gluconate IV over 10 minutes under strict cardiac monitoring. If ionized calcium levels are suggestive of hypocalcemia, the newborn should receive calcium gluconate at 8 ml/kg/d for 3 days. If seizures continue despite correction of hypocalcemia, 0.25 ml/kg of 50% magnesium sulfate should be given intramuscularly (IM).

3. Anti-epileptic drug therapy (AED)¹

Anti-epileptic drugs (AED) should be considered in the presence of even a single clinical seizure since clinical observations tend to grossly underestimate electrical seizures (diagnosed by EEG) and facilities for continuous EEG monitoring are not universally available. If aEEG is being used, eliminating all electrical seizure activity should be the goal of AED therapy.¹ AED should be given if seizures persist even after correction of hypoglycemia/ hypocalcemia (*Figure 1*).

Phenobarbitone (Pb)

It is the drug of choice in neonatal seizures. The dose is 20 mg/kg/IV slowly over 20 minutes (not faster than 1 mg/kg/min). If seizures persist after completion of this loading dose, additional doses of phenobarbitone 10 mg/kg may be used every 20-30 minutes until a total dose of 40 mg/kg has been given. The maintenance dose of Pb is 3-5 mg/kg/day in 1-2 divided doses, started 12 hours after the loading dose.

Phenytoin

Phenytoin is indicated if the maximal dose of phenobarbitone (40 mg/kg) fails to resolve seizures or earlier, if adverse effects like respiratory depression, hypotension or bradycardia ensue with phenobarbitone. The dose is 20 mg/kg IV at a rate of not more than 1 mg/kg/min under cardiac monitoring. Phenytoin should be diluted in normal saline as it is incompatible with dextrose solution. A repeat dose of 10 mg/kg may be tried in refractory seizures. The maintenance dose is 3-5 mg/kg/d (maximum of 8 mg/kg/d) in 2-4 divided doses. Oral suspension has very erratic absorption from gut in neonates, so it should be avoided. Thus only IV route is preferred in neonates and it should preferably be discontinued before discharge.

Benzodiazepines

This group of drugs may be required in up to 15-20% of neonatal seizures. The commonly used benzodiazepines are lorazepam and midazolam. Diazepam is generally avoided in neonates due to its short duration of action, narrow therapeutic index, and because of the presence of sodium benzoate as a preservative. Lorazepam is preferred over diazepam as it has a longer duration of action and results in less adverse effects (sedation and cardiovascular effects). Midazolam is faster acting than lorazepam and may be administered as an infusion. It causes less respiratory depression and sedation than lorazepam. However, when used as continuous infusion, the infant has to be monitored for respiratory depression, apnea, and bradycardia (*equipment for resuscitation and assisted ventilation should be available at the bedside of all neonates given multiple doses of AED*).

The doses of these drugs are given below:

Lorazepam: 0.05 mg/kg IV bolus over 2-5 minutes; may be repeated

Midazolam: 0.15 mg/kg IV bolus followed by infusion of 0.1 to 0.4 mg/kg/hour.

According to Volpe, the expected response of neonatal clinical seizures to anticonvulsants is 40% to the initial 20-mg/kg loading dose of phenobarbitone, 70% to a total of 40 mg/kg of Pb, 85% to a 20-mg/kg of phenytoin, and 95% to 100% to 0.05 to 0.1 mg/kg lorazepam.¹

Antiepileptic drugs for seizures refractory to above treatment

In exceptional circumstances when the seizures are refractory to the first-line AEDs, the following second-line drugs might be tried.

Maintenance anti-epileptic therapy

Principles of AED used in older children and adults are applicable to neonates also. Monotherapy is the most appropriate strategy to control seizures. Attempts should be made to stop all anti-epileptic drugs and wean the baby to only phenobarbitone at 3-5 mg/kg/day. If seizures are uncontrolled or if clinical toxicity appears, a second AED may be added. The choice may vary from phenytoin, carbamazepine, and valproic acid.

When to discontinue AED

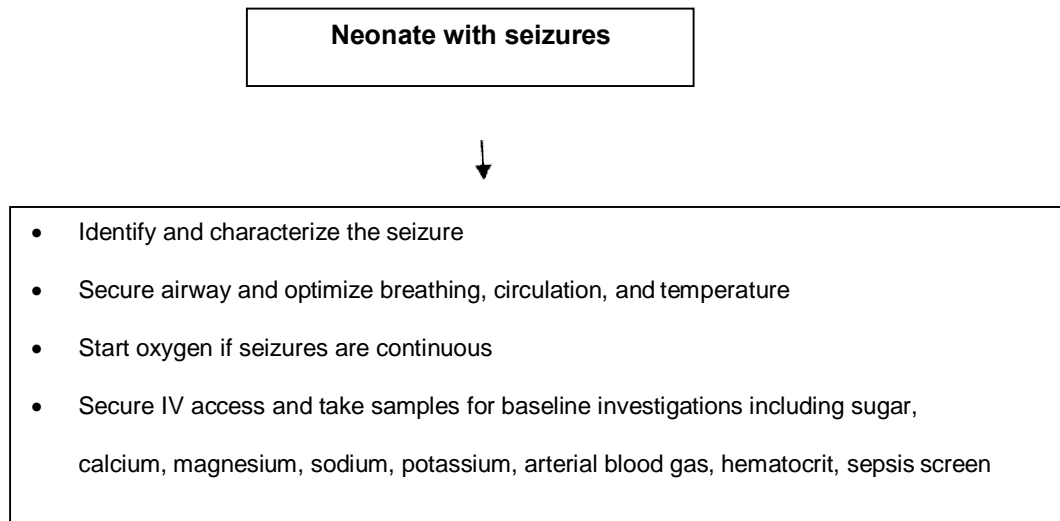
This is highly individualized and no specific guidelines are available. We follow an adaptation of the protocol recommended by Volpe.¹ We usually try to discontinue all medication at discharge if clinical examination is normal, irrespective of etiology and EEG. If neurological examination is persistently abnormal at discharge, AED is continued and the baby is reassessed at one month. If the baby is normal on examination and seizure free at 1 month, phenobarbitone is discontinued over 2 weeks. If neurological assessment is not normal, an EEG is obtained. If EEG is not overtly paroxysmal, phenobarbitone is tapered and stopped. If EEG is overtly abnormal, the infant is reassessed in the same manner at 3 months and then 3 monthly till 1 year of age (*Figure 2*). The goal is to discontinue phenobarbitone as early as possible.

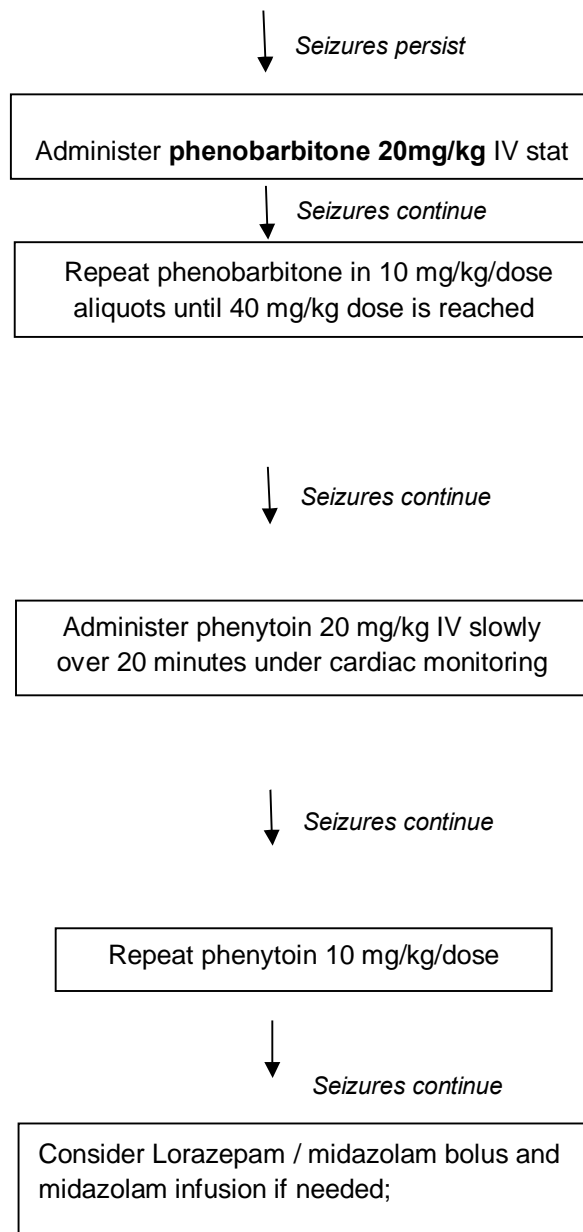
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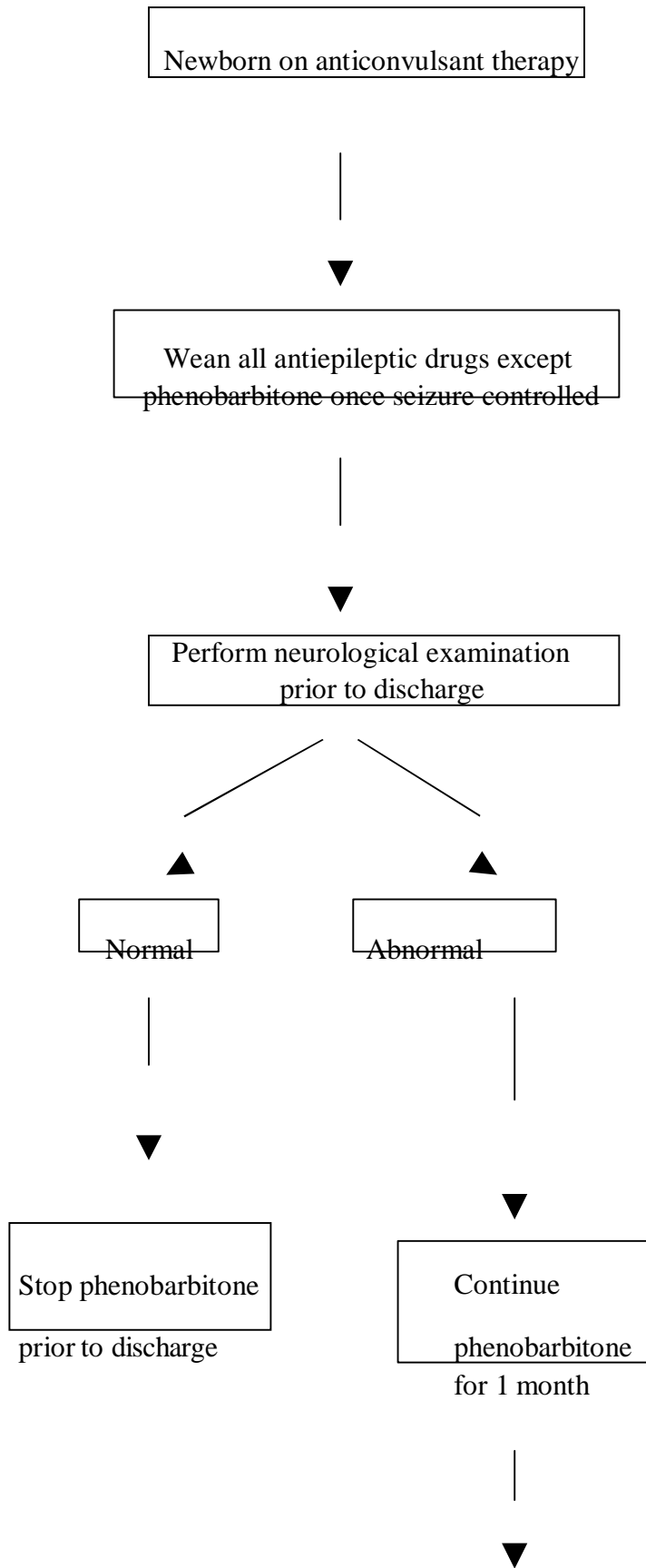
Figure 1 Acute management of neonatal seizures

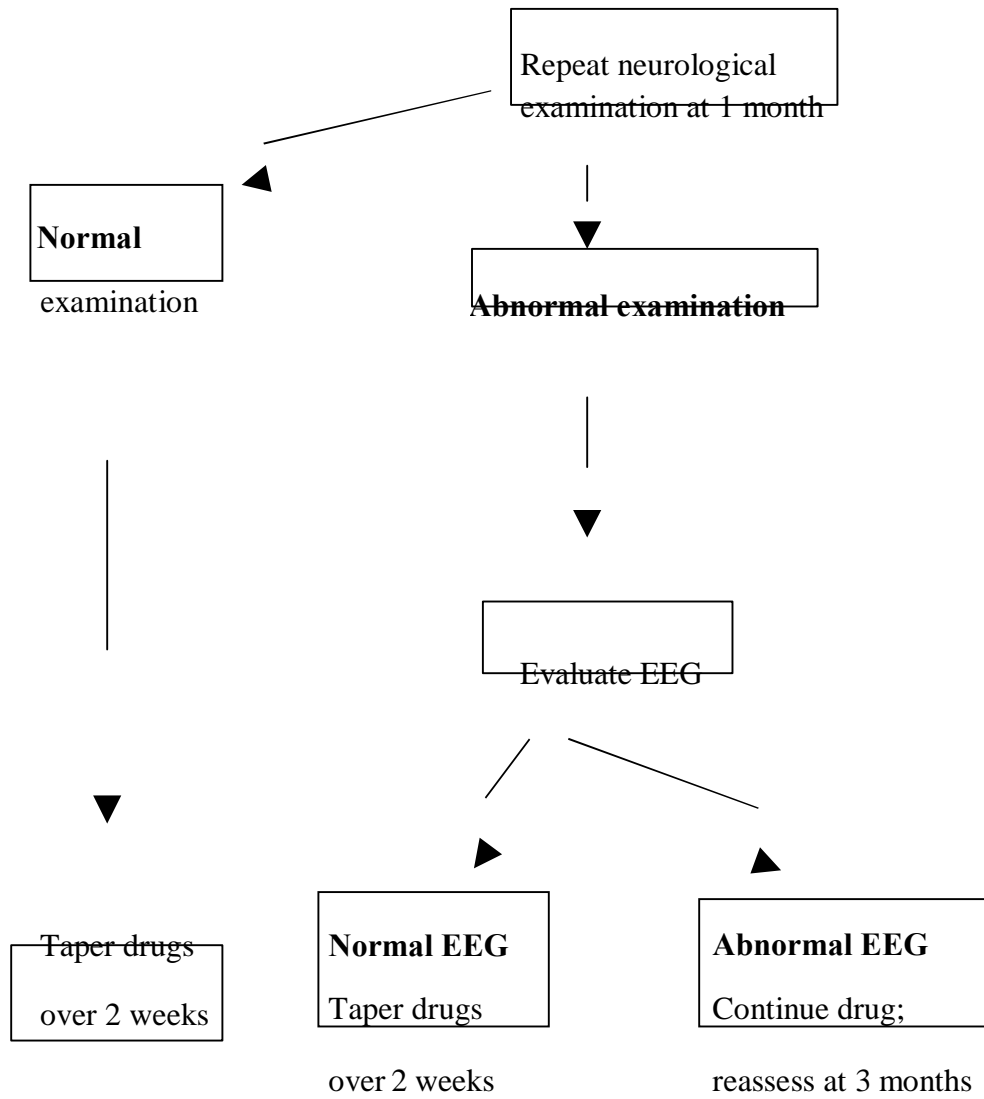




Coimbatore

Figure 2 Weaning of anticonvulsant therapy





**Intractable seizures may need lifelong therapy; consider switching over to other drugs (phenytoin or carbamazepine)*

UNDESCENDED TESTIS IN CHILDREN

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Assisted by:
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l) **WHEN TO SUSPECT/ RECOGNIZE?**

h. **Introduction :**

Diagnosis and management of the undescended testicle is required by 1 year of age as per current recommendation.

i. **Case definition:**

Cryptorchidism is the absence of one or both testes from the scrotum. It is the most common birth defect involving the male genitalia.

INCIDENCE OF THE CONDITION IN OUR COUNTRY

The incidence of cryptorchidism is 1% to 4% in full-term newborns and in up to 45% of preterm male babies. However, a large number of these will descend spontaneously by 6 months of age.

m) **DIFFERENTIAL DIAGNOSIS**

- φ. Retractable testis
- κ. Ectopic testis
- λ. Disorders of sexual differentiation (if associated with hypospadias)

n) **PREVENTION AND COUNSELING**

If a newborn boy has been found to have an undescended testis, the family needs to be counseled about the need for **review at 6 months and the possibility of surgery.**

In case of pain and swelling in the groin, there is a possibility of torsion and emergency intervention is necessary.

o) **OPTIMAL DIAGNOSTIC CRITERIA, INVESTIGATIONS, TREATMENT & REFERRAL CRITERIA**

History and Physical Examination

It is important to note if the testes were ever palpable in the scrotum at the time of birth or within the first year of life.

Classification is based on testicular location, either along the normal line of descent (abdomen, inguinal canal, external ring, pre-scrotal, upper scrotal) or in an ectopic position (usually in the superficial inguinal pouch or perineal) It is important to document associated findings such as hernia, hydrocele, penile size, and meatal position.

Check the size, location, and texture of the contralateral descended testis. Assess testicular mobility, size, consistency, and spermatic cord tension.

The key to distinguishing a retractile from an undescended testis is success of delivery and stability of the testis within the scrotum. The retractile testis will remain intrascrotal after

overstretching of the cremaster muscle, whereas a low cryptorchid testis will return to its undescended position after being released. If there is any question, a follow-up examination is indicated.

Imaging and Laboratory Tests

Routinely no imaging studies are needed. Ultrasound (US), computed tomography (CT) scans and magnetic resonance imaging (MRI) imaging studies are optional in select cases.

Basic investigations (such as hemoglobin and urinalysis) will be required for anesthetic workup.

TREATMENT

Surgery

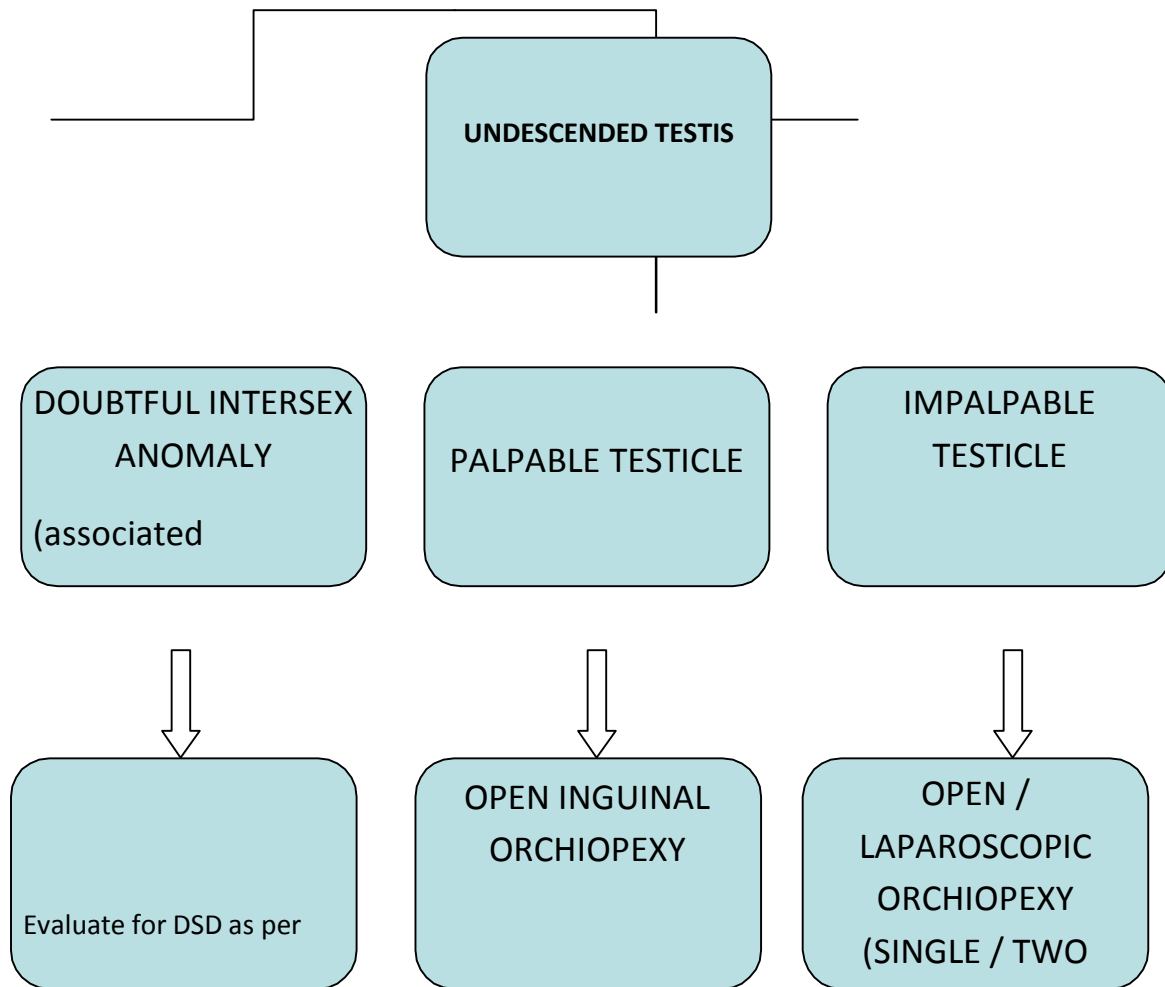
Surgery is planned between the 6th to the 12th month of age by a trained pediatric surgeon.

Palpable testis

Standard open inguinal orchidopexy is done.

Nonpalpable testis

Exploration for a non-palpable testis is usually performed with laparoscopy. Once the testes is located by laparoscopy, it may be brought down into the scrotum in a single or two stage procedure.



FOLLOW UP

Children are usually followed up as per the following time table:

10. 1 week after surgery
11. 3 months after surgery
12. Annually thereafter till puberty
13. Testicular self examination is taught to the boy at puberty

***Situation 1: At Secondary Hospital/ Non-Metro situation: Optimal Standards of Treatment in Situations where technology and resources are limited**

r. Clinical Diagnosis:

- based on history and physical examination
- it is adequate in majority of instances

s. Investigations:

- a. tests for anesthesia- Hemoglobin, urinalysis

t. Treatment:

- Surgery under general anesthesia
- Planned at 6-12months of age
- If palpable testis- standard open orchidopexy
- If non-palpable testis-laparoscopic assisted orchidopexy-either as single stage or two-stage

Standard Operating procedure

- a. In Patient: Child needs inpatient care if there are other co-morbidities that increase anesthesia risk
 - b. Out Patient: Children are evaluated and worked up as outpatients, followup too is done in the outpatient department
 - c. Day Care: Most orchidopexy operations are done as day case procedures. Children require inpatient admission if pain control is inadequate post operatively and if there are co-morbid conditions that require monitoring in the post op period.
- u. **Referral criteria:**

Child with undescended testis referred to higher centre if:

- 1.adequate anaesthesia facilities unavailable locally
- 2.intersex anomaly is suspected- as when there is:
 1. severe hypospadias with unilateral non palpable testis
 2. bilateral nonpalpable testis with or without hypospadias
- 3.child dysmorphic and a syndrome suspected

***Situation 2: At Super Specialty Facility in Metro location where higher-end technology is available**

- **Clinical Diagnosis:** as above
- **Investigations:**
 - a. for location of testis- not routinely required
 - b. for intersex anomalies;
 - i. Karyotyping,
 - ii. Ultrasonography of abdomen and pelvis
 - iii. genitography
 - iv. genitoscopy
 - v.laparoscopy

- vi. hormonal assessment
 - c. for syndromes:
 - i. genetics consultation
 - ii. karyotyping
 - iii. developmental assessment
- **Treatment:** as above
- If intersex ó as per DSD protocol
- **Standard Operating procedure**
 - a. In Patient
 - b. Out Patient
 - c. Day Care
- **Referral criteria:**
 - No further referral needed

p) ***WHO DOES WHAT? and TIMELINES***

q)

- a. Doctor makes a clinical diagnosis, counsels the family and plans surgery- a pediatric surgeon performs the surgery
- b. Nurse: assists surgeon in care of child during pre, intra and post operative course of the baby
- c. Technician: assists medical and nursing teams in care of child during intra and post-operative periods.

r) ***FURTHER READING / REFERENCES***

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**RESOURCES REQUIRED FOR ONE PATIENT / PROCEDURE (PATIENT WEIGHT
60 KGS)**

**(Units to be specified for human resources, investigations, drugs and consumables and
equipment. Quantity to also be specified)**

Situation	Human Resources	Investigations	Drugs & Consumables	Equipment
1	<ul style="list-style-type: none"> • Pediatric Surgeon • Pediatrician • Pediatric Nurse • Lab. Technician 		<ul style="list-style-type: none"> • I.V. Glucose/ Fluids • I.V. cannula • I.V. Set • anesthetic drugs, disposables • antibiotic prophylaxis 	14. Radiant Warmer 15. Saturation monitor 16. Basic Lab 17. Child friendly OT
2	<ul style="list-style-type: none"> • Pediatric surgeon • Pediatrician • Pediatric anaesthetist • Pediatric Nurse 		<ul style="list-style-type: none"> • I.V. Glucose/ Fluids • I.V. cannula • I.V. Set • anesthetic drugs, disposables • antibiotic prophylaxis 	18. ICU 19. Pediatric O.T.