GASTRO-INTESTINAL

NEC Necrotisiing Enterocolitis

BACKGROUND: This is a spectrum of disease comprising ischaemia and infection affecting the intestine of, predominantly, low birth weight, premature neonates. It may affect any part of the intestine but most commonly the terminal ileum and colon. The aetiology is unclear but it probably linked to a combination of specific bacteria in the neonatal unit and functional immaturity and impaired perfusion of the preterm infant bowel. It is very uncommon in infants who have not been fed. NEC tends to occur in sporadic clusters and affects about 2% of admissions to Neonatal Intensive Care Units. 10% of affected infants are full term. Florid cases are easily diagnosed but in many infants it is never clear if they have had "mild NEC" or not.

RISK FACTORS:
Hypoxia, neonatal asphyxia, pulmonary disease, hypotension, congenital heart disease, exchange transfusion, hyperosmolar feeding, umbilical artery/vein catheterisation, hyperviscosity syndromes (including packed cell transfusion), specific pharmacological agents and drugs (Methylxanthines{aminophylline} indomethacin, cocaine).

CLINICAL FEATURES:
The classic mode of presentation is with the triad of abdominal distension, bile stained vomiting and rectal bleeding. Any baby with these features should be assumed to have NEC and investigated accordingly. However, many preterm babies have functional gut immaturity ("slow starters") and present with distension and bilious aspirates. Some may have blood on stool testing that may be due to innocent causes (eg NG tube). Some of these babies end up being treated for NEC even though they may not have it.

General features: [Babies with NEC can be extremely ill and some die.]
Generalised sepsis: temperature instability, apnoea, metabolic acidosis, DIC, low platelets/WBC, hyponatraemia, hypotension, renal failure etc.
Abdominal features: Tenderness, distension, mass, flare, abdominal wall oedema, ascites, crepitus, inflammatory hydrocele.

INVESTIGATIONS:
All suspected cases should have FBC, film, CRP, platelet count, infection screen abdominal radiology [supine and R lateral decubitus].
The extent of other investigations will be determined by the severity of the illness but will often include blood gases, assessment of DIC, cross match etc.

X-ray findings: the classic feature is intramural gas. Other features are static or asymmetric loops on serial X-rays, ascites, portal vein gas and free air. The best way to demonstrate free air is to do a right-side-up lateral decubitus view which may show air over the right lobe of the liver. Free air can be diagnosed on a supine film (visible falciform ligament, "double contrast" effect of air on both sides of the bowel wall, liver shadow darker centrally than peripherally). Lack of free air does not exclude perforation.

MANAGEMENT:

The treatment of NEC depends on its severity.

1. Resuscitation: may need to include circulatory support (with human albumin/ blood, dopamine/dobutamine), correction of acid/base status, intubation and ventilation, platelet transfusion, WBC transfusion, immunoglobulins etc.
Most
infants with established NEC with require analgesia with morphine infusion.

2. All babies with suspected or proven NEC should be commenced on:
Nil by mouth]
Intravenous feeding] for 10 days
Antibiotics]
Tazocin
Vancomycin
Metronidazole 7.5 mg/kg 3 times a day

3. Any "at risk" factors (see above) should be avoided.

4. A nasogastric tube must be passed and kept on free drainage with hourly aspiration.

5. Regular repeated assessment both clinically and radiologically. The frequency of assessment will depend on the state of the child but should include, initially, <u>at least</u> daily haematology, biochemistry.
Decubitous views should be repeated if the bowel gas pattern changes or the clinical condition deteriorates.

6. Surgery: Usual indications for emergency surgery are - perforation
- dead bowel
Assessment of the need for, and timing of, surgery may be difficult. Free air is usually an indication for surgery although in very small babies (eg <1000g) surgery may be avoided and a peritoneal drain inserted instead. Dead bowel is usually manifested by an abdominal mass, erythema and persisting acidosis/thrombocytopoenia/hyponatraemia. Surgical treatment will comprise excision of any dead gut with either primary anastamosis or stoma formation [or open and close with relook at 48 hours if all gut affected].

LATE COMPLICATIONS

Most babies with mild NEC make a complete recovery. More severe cases may develop intestinal strictures in the small or large bowel or both. These usually present with distension and feed intolerance up to 6-8 weeks after the acute illness and can usually be detected by contrast studies. Extensive bowel resection may result in short bowel syndrome and long term TPN.

Congenital Diaphragmatic Hernia

Presentation: the timing of presentation depends on the severity of lung hypoplasia with

3 main clinical patterns:

1. Severe respiratory distress at birth never achieving oxygenation
2. Respiratory distress in the first few hours of life
3. Mild respiratory distress inthe first few days of life

In addition many babies are now diagnosed prenatally on ultrasound scan.

Diagnosis: 80% of CDH are left sided.

Clinical scaphoid abdomen
mediastinal shift to R usually [->apex and trachea]
bowel sounds in the chest

Radiological CXR bowel loops in chest
mediastinal displacement
AXR paucity of bowel loops in abdomen

Differential diagnosis: congenital adenomatiod malformation of the lung [CCAM]. It is essential to perform an AXR in all cases of suspected CDH to demonstrate a paucity of bowel loops in the abdomen and thus exclude CCAM.

Associated anomalies: found in 40% of liveborn CDH (neural tube, cardiac, skeletal, renal etc). Malrotation is present to some degree in all cases.

Management prior to transfer for surgery:

All infants will require: CXR and AXR
blood gas, FBC, U&E, X-match
pass NG tube - free drainage + hourly aspiration

Respiratory management depends on the severity of lung hypoplasia. A good clue to the severity of this is the timing of presentation with respiratory symptoms. If this is after 2 hours of age survival approaches 100% and persistent pulmonary hypertension of the newborn [PPHN] occurs in 20% of cases. Presentation before 2 hours is associated with a >60% mortality and 90% develop PPHN incidence.

Presentation <2 hrs old or after prenatal diagnosis

Prenatal management
- fetal karyotyping
- prenatal counselling by Neonatologist and Paediatric Surgeon
- in utero transfer to RSCH for delivery
- Dexamethasone prenatally may reduce postnatal respiratory distress

At delivery - elective intubation at birth
- DO NOT VENTILATE WITH BAG AND MASK as this inflates the intrathoracic

stomach and compromises lung expansion
- intubation
- give surfactant 100 mg/kg Curosurf
- IPPV: aim for PaO_2 10-12 kpa, $PaCO_2$ 3.5-4.5 kpa, pH 7.45-7.55
- paralyse [pancuronium]
- sedate [morphine]
- arterial line [in a preductal vessel eg R radial if possible, or high UAC]
- plasma 10-20 ml/kg to maintain bp
- pre and post ductal SaO_2 +/- TcO_2 monitoring
- transfer baby when stable to UNIT

Stabilisation on UNIT
- nurse under overhead heater
- fluid restrict to 70 ml/kg/d initially
- double lumen central line via RIJV
- dopamine at 5 mcg/kg/hr via central line to improve tissue/renal perfusion
- (dobutamine if needed for inotropic effect on bp)
- pass urinary catheter
- moniter arterial pressure and CVP. Ideal CVP 2-3 mm Hh above mean airway pressure
- TPN to start when convenient (NG feeds may be possible in due course)
- arrange echocardiography - to exclude congenital anomaly and document PPHN
- vasodilator management of PPHN as per protocol if required

Surgery is only undertaken when stable in FiO_2<40% if this is ever achieved.

<u>Presentation > 2 hrs of age</u>

Prior to transfer for surgery
- DO NOT VENTILATE WITH BAG AND MASK as above
- intubation only if required, with standard IPPV
- arterial line [preductal if possible or UAC]
- plasma 10-20 ml/kg to maintain bp
- pre and post ductal SaO_2 +/-TcO_2 monitoring
- transfer within hours

After transfer
- nurse in incubator
- surgery in first 24-48 hours unless PHT develops requiring treatment as above

<u>Surgery</u>
- transfer to theatre under O/H heater
- patient to be accompanied throughout theatre visit by the nurse specialing for that shift
- all preop treatment to be continue peroperatively

Surgical repair is usually by primary closure although a prosthetic patch repair may be required in severe cases. Chest drains are not routinely inserted. Malrotation may require correction.

<u>Postoperative care</u>
- continue as per pre op
-anticipate worsening PHT after surgery
- mediastinal shift:

centralisation of the mediastinum using a balloon catheter chest drain or by injecting air into the ipsilateral pleural space may be required

- pleural effusion:

this always develops over some days postop to fill the void left by the hernia. It is rarely under tension and rarely requires drainage.

ECMO [extracorpreal membrane oxygenation]

This form of heart lung bypass may buy the baby time whilst the pulmonary vasculature matures if the baby has sufficient lung tissue. A good indication of adequate lung development is achieving a normal PCO_2.

ECMO inclusion criteria:>35 weeks completed weeks gestation at birth, >2kg, <8 days IPPV, <28 days old. Oxygenation index (0I)>35 for 3 or more hours is an indication to discuss with GOS. 0I>40 = ECMO or $PaCO_2$ >12 kpa for 3 or more hours.

$$0I = \frac{\text{mean airway pressure} \times FiO_2}{PaO_2} \times 100$$

Background

CDH has an incidence of 1:2000 fetuses but, because of associated chromosomal anomalies [trisomy 13/18] and other severe congenital anomalies, approx 1/3 of fetuses die in utero. In liveborn babies the incidence is about 1:3000. CDH has a high mortality [40-60%] which is largely due to lung hypoplasia which usually affects both lungs. In some babies this is so severe that adequate oxygenation is never achieved and death occurs within hours of birth. At the other extreme some babies present with minor respiratory symptoms after 1-2 days of life and have essentially normal lungs. However, many have enough lung to become oxygenated but face severe problems from pulmonary hypertension.

Rational for above management

The pathology of pulmonary hypoplasia includes muscular hypertrophy of pulmonary arterioles. These vessels produce a high pulmonary vascular resistance and right-to-left shunting at atrial and ductal levels (persistant fetal circulation PFC). They are very prone to spasm which will worsen the shunt and may be induced by such stimuli as pain, cold, hypotension, acidosis, hypoxia etc. Once established PFC may be hard, if not impossible to reverse. Paralysis, sedation etc may help prevent PFC worsening. Hyperoxia > 12 kpa, hypocarbia = 4 kpa and alkalosis pH > 7.5 all help stabilise pulmonary arterioles. The tendence of pulmonary arterioles to spasm should decrease with time and the risk of PFC should diminish. Surgery is often followed by clinical deterioration, partly due to the trauma of surgery and partly due to postoperative mediastinal shift towards the side of the hernia, overinflation of the only good lung (usually the right) and worsening lung compliance. For this reason surgery is delayed until there are signs that the pulmonary vasculature is stable enough to withstand the insult of surgery. Many CDH babies who develop PFC require the above management for some days prior to attaining FiO_2 of 40% required for surgery and some will die in the attempt. Many will become grossly oedematous due to paralysis hence the need for early fluid restriction. If all else fails vasodilators such as tolazoline may help but most drop the systemic blood pressure which must be normalised using plasma or inotropes beforehand.

Outcome

Mortality	overall liveborn	50%
	liveborn presenting <2 hrs	60%+
	" " >2 hrs	0%
	overall fetus	85%
	fetus with isolated CDH	60%

Long term complications

- chronic lung disease: bronchopulmonary dysplasia may occur in survivors
- gastro-oesophageal reflux: may be severe and associated with poor feeding requiring gastrostomy
- thoracic cage deformities: may develop in early childhood