Neonatal Intestinal Failure (including Short Gut)

Overview and definition

Intestinal failure (IF) can be defined as “a reduction in the functioning intestinal mass below the amount necessary for adequate absorption to allow growth”. Within this group there are babies with short gut, motility disorders and primary mucosal disorders. Initial survival will depend on receiving Parenteral Nutrition (PN). In the neonate, the largest contributing factor to IF is short gut (often following surgical resection) secondary to Nectrotising Enterocolitis (NEC), Gastrochisis or small bowel atresia (or a combination of all three). Neonates with IF suffer increased sepsis, cholestasis, central venous cannula problems, have longer admissions, may develop PN associated liver disease and increased morbidity.

Post surgical prognosis depends on the residual length and type of the small bowel, the presence of the ileo-caecal valve (IC) and whether or not the colon has been preserved. The IC valve is said to act as a barrier to colonic bacteria and its loss may lead to bacterial overgrowth. It also “brakes” the exit of fluid and nutrients from the small intestine, loss of the IC valve can lead to diarrhoea initially.

At 28 weeks, the small bowel is estimated to be 150cm long and, by term, has grown to around 240cm in length. The small intestine has a large functional reserve and can tolerate 40-50% resection without major (long term) nutritional sequelae. The minimum length of gut required to avoid short gut related IF is impossible to define. However, with an IC valve and intact colon, as little as 30cm may be enough but without an IC valve a longer length is usually needed to avoid long term dependence on PN. Estimating bowel length is difficult and it is equally important to consider the type of bowel (i.e jejunum or ileum) as well as its residual motility. Jejunum is more important overall but ileum has the greater capacity for adaptation.

Intestinal adaptation starts to occur 24-48 hours after resection with epithelial hyperplasia, increased length of villi and the increased surface area resulting in improved digestive and absorptive abilities. Animal studies suggest that the mucosal hyperplasia requires direct enteral nutrition.

Parenteral Nutrition Associated Liver Disease (PNALD)

PNALD develops in 40-60% of infants requiring long term PN. It is reversible provided enteral feeding is established, PN kept to a minimum and sepsis avoided.

Several causes are postulated including: poor bile secretion / stasis secondary to fasting, excessive glucose administration in PN leading to fatty liver or hepatic fibrosis, excessive protein intake in PN leading to reduced bile flow and phytosterols in PN producing direct oxidant damage to the liver. However, most significant is recurrent septicaemia which many infants suffer while receiving long term PN. This may be caused by bacterial overgrowth in poorly motile bowel or the close proximity of stomas to tunnelled central lines.

Jaundice correlates with morbidity & mortality. Aggressive enteral feeding has been shown to lead to resolution of jaundice and improved transaminase and alkaline phosphatase levels. Repeated infections increase bilirubin levels and worsen prognosis.

Surgical Management

When possible stomas should be closed to avoid sepsis, if stomas are necessary then distal loops should be fed. Bowel lengthening surgery may be indicated (the Bianchi technique or STEP procedure) which can lead to lengthening of bowel and increasing surface area resulting in the successful weaning off PN. Timing of surgery and where surgery should take place must be considered carefully. Intestinal transplantation is an emerging possibility that may be considered for irreversible PN dependent intestinal failure usually in the face of progressive liver failure. Due to advances in immunosuppression, long term outcomes of transplantation are improving and patients report greater quality of life post transplantation. In the Birmingham Children Hospital, the overall 5 year survival following small bowel transplantation was previously reported to be around 50%. However, this figure is probably far higher by now but organ availability and technical difficulties make this difficult in infants <10kg. To date, two Welsh children have been transplanted (1 small bowel transplant who died of post transplant lymphoproliferative disease and 1 combined liver and small bowel who is doing well 2 years post transplant).

Management Principles

Within a month, all cases of neonatal IF should be discussed in detail between the Neonatologist, Surgeon and a Gastroenterologist. Ideally, they should then be reviewed weekly by the Nutrition team. The priority is to attain and maintain growth and development whilst transferring the feeding from the parenteral to the enteral route. Try to avoid changing more than one aspect of the management at the same time.
Feeding
This is crucial as aggressive enteral feeding can reverse PNALD.

- Breast Milk is preferred but Peptijunior is often considered because it is glucose based, hydrolysed, high in MCT fat and has a lower osmolality than amino acid formula.
- Wherever possible, the oral route should be utilised and early weaning may be considered. Even very small volumes of oral feeding can be beneficial and the early involvement of Speech and Language Therapists is advisable.
- Continuous nasogastric feeds improve feed tolerance.
- It is often necessary to tailor the regime to the baby e.g. some oral or bolus daytime feed e.g. EBM or Cows Milk Formula with an overnight nasogastric infusion of Peptijunior.

Medications to be considered
- Loperamide can be used to slow gut transit times.
- Ranitidine may be helpful if diarrhoea is problematic, this is often due to gastric hyper section. If the jejunum has been removed there is loss of negative feedback for gastrin. Omeprazole may also be considered and is sometimes used in addition to Ranitidine.
  - Ursodeoxycholic Acid is started when conjugated bilirubin is > 50mmol/l. It improves bile flow and reduces gall bladder and intestinal stasis, has minimal side effects.
  - Cholestyramine can bind bile acids and decrease diarrhoea. It may also reduce oxalate absorption which would decrease stone formation.
  - Enteral antibiotics (metronidazole, gentamicin or vancomycin) if bacterial overgrowth is a concern.

Parenteral Nutrition not Total Parenteral Nutrition
Long term Parenteral nutrition (PN) should be infused via a single lumen catheter with its tip positioned in either the superior vena cava or inferior vena cava in order to decrease the risk of infection and thrombosis. Local preference is for a tunneled Central line to be considered if an infant is likely to need long term PN support and transfer to the paediatric wards. This is secure, allows PN to be concentrated and given over shorter infusion times.
It is imperative to reduce the risk of line infection as each infection can lead to a >30% increase in bilirubin levels and will contribute to the progression of liver disease. Preventative measures such as strict aseptic techniques and avoiding blood sampling from central lines are vital.
If a line infection is suspected, take blood cultures peripherally and from the central line and start antibiotics. If the patient remains unwell despite antibiotics, the line may still be infected and will need to be removed. First line antibiotics are Vancomycin and Gentamicin. (If there are concerns regarding a persistent infection, consider changing to Teicoplanin and Gentamicin as can be administered once daily via the tunneled central line). Traditionally used lipid emulsions containing omega-6 PUFAs originating from plant oils may cause liver injury. Moreover, newer lipid emulsions containing fish oils eg SMOF reverse jaundice within 9 weeks of use. Accordingly, SMOF (Soya MCT, Olive and Fish Oils) is now used as the lipid infusion of choice on the NICU. In neonates awaiting transfer to the paediatric wards consider concentrating PN to less than 100 ml/Kg/day, try to reduce infusion times below 24 hours, give “lipid holidays” and give sufficient calories to ensure growth of 140-210 g per week. Feed closer to expected weight than current weight. Beware of giving too much Calcium to term babies and remember PN is relatively deficient in Iron.

Monitoring
Bloods should be checked as per UHW NICU PN guideline.
Urinary Sodium should be checked weekly, aim for greater than 20mmol/l.
Weigh at least twice a week, head Circumference and length (when possible) should be plotted weekly. Aim for 140 – 210g weight increase per week.
Monthly monitoring of vitamins A, D &E, Trace elements, B12 and folate is recommended.
After 2 months on PN consider performing an Abdominal and Renal USS, looking for hepatic echogenicity, vessel flow, splenic size and presence of renal stones.

References
Stringer and Puntis. Short Bowel Syndrome. Archives of Disease in Childhood 1995; 73, 170-173

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