nutrition and preventing oesophageal stricture. Earlier controversies regarding steroid treatment in preventing stricture formation have been evaluated in a number of recent studies.[13–15] Anderson *et al* showed that with first and second degree oesophageal injuries, strictures developed in 5% of the steroid treated patients and none of the controls[13]; with third degree injuries, strictures occurred in 90% of the steroid-treated children and 100% of the controls. Stricture formation is thus related to the degree of burn irrespective of steroid treatment.

In the absence of oesophageal or gastric perforation, the use of antibiotics has not been clarified. Animal studies have shown that the oesophageal burn creates a portal of entry for bacteria. There is, however, insufficient data to support the routine use of prophylactic antibiotics.

The use of a nasogastric tube in caustic injury is also debatable. [16 17] If employed it should only be passed after the degree and severity of caustic injury has been assessed at endoscopy. The indications for nasogastric intubation are for nutritional purposes and to function as an intraluminal stent.

Oral feeding in caustic injury is commenced as soon as the patient is able to swallow saliva. In patients with dysphagia an oesophagogram using water soluble contract material and oesophagoscopy should be performed 7–10 days after the acute injury to evaluate the extent of damage. If a stricture is demonstrated, bougienage is commenced three weeks later using prograde dilatations or retrograde dilatations via a gastrostomy and the placement of a transoesophageal 'string'. If gastro-oesophageal reflux is demonstrated in the presence of persistent stricturing antireflux surgery is indicated.[18]

Where dilatations have failed or when the oesophagus has been damaged beyond salvage, the choice of the ideal substitution remains open to debate. The operations currently in use are jejunal interposition,[19] gastric tube oesophagoplasty,[20] colon interposition,[21–22] and gastric transposition.[23]

Risks of cancer

The incidence of cancer after corrosive stricture of the oesophagus is less than 5% and only occurs after a long latent period.[24] Gerzic *et al* report an incidence of 8% of cancer following caustic injury to the oesophagus 25 to 50 years previously.[25] This association raises the importance of the need for long term follow up and may influence the decision to resect and replace the injured oesophagus.[26] The mortality risk for oesophagectomy (11%)[11] by far surpasses the risk of cancer in the scarred oesophagus.[27] A selective approach to oesophagectomy is advocated. Oesophagectomy is performed only if there is a complete stricture of the thoracic oesophagus. Long term follow up is crucial in oesophageal caustic injury in view of the risk of cancer.

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Failure to thrive in congenital heart disease

'The children rarely thrive and often display a lethargy of mind and body'

Osler on the symptoms of congenital heart disease[1]

The outlook for congenital heart disease (CHD) has changed enormously since Osler's day, although failure to thrive remains a common presentation. How common is not precisely known as the natural history of CHD changes constantly as medical and surgical opportunities for treatment expand.[2] The study often quoted as evidence for the frequency of failure to thrive in CHD, reviewed children over a wide age range 30 years ago and has long been superseded.[3] Patent ductus arteriosus for example, cited as one cause of prolonged failure to thrive,[3] is now usually corrected shortly after diagnosis and rarely causes significant growth problems. Thus our knowledge about the frequency and causes of failure to thrive in CHD needs updating. Infants at particular risk should be recognisable so specific

nutritional management can be instituted before failure to thrive intervenes.

Why do some infants with CHD fail to thrive?

There is a variety of reasons why infants with CHD fail to thrive.[4 5] CHD is occasionally part of syndromes,[6] which include intrauterine, and/or postnatal, growth retardation (for example congenital rubella, Turner's syndrome, fetal alcohol syndrome). A low energy intake in some infants is another reason for failure to thrive. Anorexia is a recognised symptom of cardiac failure in adults[7 8] and anorexia, breathlessness, or anoxia can prevent infants ingesting and retaining the volumes of feed necessary to meet nutrient requirements. Hypochloraemic alkalosis secondary to vigorous diuretic treatment may be another explanation for anorexia.[5] Perhaps unexpectedly, malabsorption secondary to anoxia or bowel wall congestion is rarely a cause of failure to thrive in CHD.[9 10] Sondheimer and Hamilton studying CHD infants more than 20 weeks old found increased fat loss (>15% stool fat) in five out of 21 infants and increased stool protein losses in nine out of 15 infants.[9] Other studies have found less evidence for malabsorption.[11] We did not find significantly increased stool energy losses in any CHD infants compared with control infants.[10]

Infants at particular risk of failure to thrive are those with cyanotic CHD and those with left to right shunts, pulmonary hypertension, and right sided cardiac failure. With cyanosed infants, the severity of failure to thrive is not necessarily proportional to the degree of cyanosis. It is not clear why, as hypoxia, acidosis, and consequent anaerobic metabolism influence cardiac efficiency. In the mammalian fetus, carbohydrates are the major myocardial substrate but early in postnatal life the heart switches from a predominantly carbohydrate metabolism to deriving two thirds of energy from β-oxidation of free fatty acids.[12] During hypoxia the heart switches back to glycolytic metabolism. Unmetabolised fatty acids then exert inhibitory effects on carbohydrate metabolism and are a cause of decline in myocardial contractility. Metabolic inefficiency of the heart and other tissues contributes to failure to thrive by allowing little spare energy for growth.

Infants with failure to thrive, significant left to right shunts, and pulmonary hypertension show increased resting energy consumption (measured indirectly by oxygen uptake, $\dot{V}O_2$), compared with body weight.[10] These infants are usually grossly underweight. As lean tissue has higher resting metabolic rate than fat, the high resting metabolic rates may be artefacts related to the altered body composition of these children. This would not explain the initial failure to thrive despite apparently adequate energy intakes.[13–16] Comparing CHD and control infants, we found resting $\dot{V}O_2$ showed similar correlations with weight for age and summed skinfold thickness.[10] However the control children were growing very well and many of the CHD children were not growing at all. CHD children had little spare energy for growth.[14] Clinical features of pyrexia and profuse sweating in many severely affected CHD children with failure to thrive support the concept of increased resting metabolism/kg lean body mass in at least some children.

There are good reasons for expecting resting $\dot{V}O_2$/lean body mass to be increased in some children with CHD. Oxygen consumption of the heart in normal adults requires about 11% of total energy requirements but in severe aortic stenosis, cardiac oxygen consumption may increase to 32% of total energy requirements.[7 8] Eighty percent of energy consumed by the mature heart is used in pumping activity and only 20% in basal metabolism and electrical activity.[12] Most children with CHD and severe wasting have pulmonary hypertension. Greatly increased oxygen requirement by a hypertrophied right ventricle working against high resistance could be the energy demanding process.

Gingell and Hornung have compared height, weight, and weight for height in infants with tetralogy of Fallot (TOF) and with ventricular septal defect (VSD).[5] Both groups of children were on average short for age but weight for age was lower in the children with VSD and weight for height quite markedly lower than for those with TOF. Children with TOF were stunted whereas those with VSD were wasted.[5] These differences in nutrition would suggest rather different aetiologies for growth retardation and different potential for catch up growth on correction of the cardiac defects.[17] Catch up growth is more dramatic with wasting than stunting and the children with VSD showed accelerated weight gain after surgery whereas growth of those with TOF was relatively unchanged. Failure of the children with TOF to catch up has not been found in all studies.[18] Categorisation of the different growth potential in CHD—with or without failure to thrive—should help indicate likely growth and nutritional outcomes from surgery. This could be useful in planning management.

How can we improve the growth of children with CHD?

We need to recognise the diagnostic groups at particular risk of failure to thrive and direct efforts towards ensuring these infants achieve at least normal nutrient recommendations in early life. This may require replacing vomited feeds, spending time feeding infants who are slow, or providing nasogastric gavage for reluctant feeders. Continuous slow nasogastric gavage feeding has been effective in some infants.[11 19 20] Supplementation with glucose polymers will increase energy intakes at lower volumes of intake and encourage growth. It may be logical to use carbohydrates as a source of extra energy rather than fat or protein as shifting cardiac metabolism from β-oxidation of fatty acids to carbohydrate utilisation has beneficial effects even in mature stressed hearts.[12]

High energy feeding by gavage should be introduced only if other methods seem to be failing. While it makes it easier to feed the volumes of supplemented formula necessary for growth, it provides no normal appetite experience and no education in feeding skills. Children who have had major pathological problems with feeding in early infancy often show major behavioural problems with feeding once the pathology has resolved.[21] Failure to thrive may be prevented in early infancy only for weanlings to refuse to feed later.

When infants fail to thrive despite these measures, there should be a reconsideration of the best time for corrective surgery. Although the risks increase with deterioration in nutritional state, surgery may be the only reasonable option for a child with progressive failure to thrive.[18] The child may be small and malnourished now but, in the absence of normal growth, the severity of malnutrition can only increase with time. Moreover, infants with no spare energy for activity miss the experiences so important to normal developmental progress.

Little is written about the neurodevelopmental outcome in CHD.[22] Preoperative neurological assessment is rarely recorded and most studies attribute later neurodevelopmental problems to the complications of surgery.[22] However, infants with transposition of the great vessels show lower neurodevelopmental scores with longer waits for surgical correction.[23] Preoperative anoxia, malnutrition, and stimulus deprivation could all act to delay developmental progress in many children with CHD.

Developmental delay in CHD has as many origins as failure to thrive. Whatever the cause, it is undesirable. Management of nutritionally stressed infants with CHD should therefore include programmes for positive stimulation

for these infants to recompense for their restricted environment. Feeding the starved mind may, in the long term, be as important as feeding the starved body.

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Renal stone disease—investigative aspects

In Europe, renal stone disease affects between one and two children/million total population/year.[1] The disease is more prevalent in less industrialised countries where a high proportion of bladder stones are found.[2] Most patients present in the early years of childhood,[3] usually with a urinary tract infection, abdominal pain, or haematuria.[4] Although the majority have only one episode of stone formation,[5] it is important to identify risk factors wherever possible because specific treatments can reduce stone recurrence and renal damage in those most at risk.[6 7]

Aetiology

Renal stones result from the precipitation and growth of crystals within the urinary tract. Precipitation may be encouraged by an increased concentration of insoluble materials, a change in pH,[6] or a reduction in concentration of one or more of the physiological inhibitors of crystal growth. The most important inhibitors are mucopolysaccarides, citrate, and pyrophosphate.[8 9] It is more difficult for crystals to form spontaneously than for material to precipitate on pre-existing crystals or other solid matter. This gives rise to the phenomenon of epitaxy where there may be an aggregation of material that is not necessarily the same as that of the original nidus.[6]

A variety of clinical disturbances can result in one or more of these conditions.

INFECTION

Within Europe the commonest identifiable factor is infection with urease producing organisms, proteus accounting for over 80% of cases.[3 4 10] Infection related urolithiasis is less common in the USA, where there is an overall lower incidence of stone disease.[11] The likely mechanism of stone formation is that ammonia, liberated from the action of urease on urea, increases urinary pH, decreasing the solubility of calcium salts and increasing the concentration of the ammonium ion which can precipitate with phosphate and magnesium (struvite). The majority of infection related stones consist of struvite with lesser amounts of calcium apatite (calcium hydrogen phosphate)[7] and matrix material.

ANATOMIC ABNORMALITIES

Urolithiasis is associated with those developmental abnormalities of the urinary tract that result in urine stasis and lead to a greater risk of infection. These stones are predominantly calcium phosphate together with struvite.[10]

METABOLIC STONES

Children in whom a metabolic defect has been demonstrated are in the minority, 10 to 20% in Europe.[10 11] However this may well be an underestimate because not all children receive a metabolic evaluation.[12] The majority of those reported have idiopathic hypercalciuria,[13] associated either with enhanced gut absorption, increased renal excretion, or enhanced mobilisation of calcium from bones as commonly occurs during prolonged immobilisation. Distal renal tubular acidosis can also lead to the formation of calcium stones. The combination of an increased urine pH, low citrate, and high calcium excretion enhances calcium precipitation.[7] Diseases associated with hypercalcaemia rarely present with renal stones; they are more likely to lead to nephrocalcinosis.

The next most common metabolic cause of stones is cystinuria accounting for up to 2% of cases of urolithiasis.[14] Cystinuria is a consequence of defective renal reabsorption of basic amino acids resulting in urinary concentrations of cystine that exceed its solubility. However not all individuals with the defect form stones, and not all the stones formed are pure cystine.[14]

Urate crystaluria and occasionally urolithiasis may be a complication of haematological tumours after chemotherapy. Hyperuricaemia may also be caused by a deficient activity of the enzyme hypoxanthine-guanine phosphoribosyl transferase (HGPRT). Complete deficiency gives rise to Lesch-Nyhan disease but partial defects have also been described.[15]

A deficiency of the next enzyme in the same pathway, xanthine oxidase, is responsible for the systemic accumula-