Indications for transoesophageal studies in children

- (1) **Primary diagnosis:**
 - Atrial pathology
 - Anomalous venous connections
 - Atrial septal defects
 - Atrioventricular junction abnormalities
 - Left ventricular outflow obstruction
- (2) **Monitoring:**
 - Perioperative
 - Atrial lesions
 - Atrioventricular valve repair/replacement
 - Mustard/Senning procedure
 - Fontan procedures
 - Continuous monitoring
 - Early postoperative complications
 - Interventional cardiac catheterisation
 - Balloon valvuloplasties
 - Balloon angioplasties
 - Atrial septostomies
 - Venous pathway dilatations
 - Coil embolisations
 - Atrial septal defect occlusion
 - Ventricular septal defect occlusion
- (3) **Follow up:**
 - Venous pathway reconstructions
 - Atrioventricular valve repair/replacement
 - Mustard/Senning procedures
 - Fontan procedures

lesions. After the Mustard or Senning procedure for complete transposition, transoesophageal studies allow a complete evaluation of atrial baffle function,¹² and thus largely influence the need for follow up cardiac catheterisation. After the various modifications of the Fontan procedure, which is used for palliation of a range of complex congenital cardiac lesions, the transoesophageal ultrasound approach provides a most sensitive tool in the definition of residual or acquired haemodynamic lesions and yields new insights into the patterns of the pulmonary circulation.¹³

Limitations

Paediatric transoesophageal echocardiography is a semi-invasive technique that requires either general anaesthesia or heavy sedation. Thus appropriate patient selection is mandatory (table). With the currently available dedicated paediatric scanning equipment studies in children below 4 kg in weight should be performed only if the information likely to be obtained is of importance for appropriate patient management. The present probe technology allows for only single plane imaging of the heart, thus making transoesophageal scanning a strictly tomographic technique. However, it is likely that biplane^{14 15} or multiplane paediatric transoesophageal probes will be designed in the near

future and will further increase the range of diagnostic insights to be obtained.

Conclusion

Transoesophageal echocardiography is a relatively new adjunct to the ultrasound evaluation of congenital cardiac lesions. It is a safe, albeit semi-invasive, technique that rapidly has gained a well defined place in the diagnostic armamentarium of the paediatric cardiologist.

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- 1 Gussenhoven EJ, Taams MA, Roelandt JRTC, *et al.* Transoesophageal two-dimensional echocardiography: its role in solving clinical problems. *J Am Coll Cardiol* 1986;8:975-9.
- 2 Kyo S, Koike K, Takanawa E, *et al.* Impact of transoesophageal Doppler echocardiography on pediatric cardiac surgery. *Int J Card Imaging* 1989;4: 41-2.
- 3 Ritter Sb, Hillel Z, Narang J, Lewis D, Thys D. Transoesophageal real time Doppler flow imaging in congenital heart disease: experience with the new pediatric transducer probe. *Dynamic Cardiovascular Imaging* 1989;2:92-6.
- 4 Stümper O, Elzenga NJ, Hess J, Sutherland GR. Transoesophageal echocardiography in children with congenital heart disease—an initial experience. *J Am Coll Cardiol* 1990;16:433-41.
- 5 Stümper O, Sreeram N, Elzenga NJ, Sutherland GR. The diagnosis of atrial situs by transoesophageal echocardiography. *J Am Coll Cardiol* 1990;16: 442-6.
- 6 Stümper O, Rijlaarsdam M, Vargas-Barron J, Romero A, Hess J, Sutherland GR. The assessment of juxtaposed atrial appendages by transoesophageal echocardiography. *Int J Cardiol* 1990;29:365-71.
- 7 Stümper O, Vargas-Barron J, Rijlaarsdam M, *et al.* The assessment of anomalous systemic and pulmonary venous connections by paediatric transoesophageal echocardiography. *J Am Coll Cardiol* 1991;17:256A.
- 8 Sreeram N, Stümper O, Kaulitz R, *et al.* The comparative value of surface and transoesophageal ultrasound in the assessment of congenital abnormalities of the atrioventricular junction. *J Am Coll Cardiol* 1990;16:1205-14.
- 9 Roberson D, Muhiudeen I, Silverman N, Turley K, Cahalan M. Intraoperative transoesophageal echocardiography in infants and children with cardiac shunt lesions. *J Am Soc Echocardiogr* 1990;3:213.
- 10 Stümper O, Kaulitz R, Sreeram N, *et al.* Intraoperative transoesophageal versus epicardial ultrasound in surgery for congenital heart disease. *J Am Soc Echocardiogr* 1990;3:392-401.
- 11 Hellenbrand WE, Fahey JT, McGowan FX, Weltin GG, Kleinman CS. Transoesophageal echocardiographic guidance of transcatheter closure of atrial septal defect. *Am J Cardiol* 1990;66:207-13.
- 12 Kaulitz R, Stümper O, Geuskens R, *et al.* The comparative values of the precordial and transoesophageal approaches in the ultrasound evaluation of atrial baffle function following an atrial correction procedure. *J Am Coll Cardiol* 1990;16:686-94.
- 13 Stümper O, Sutherland GR, Geuskens R, Roelandt JRTC, Bos E, Hess J. Transoesophageal echocardiography in evaluation and management of the Fontan circulation. *J Am Coll Cardiol* 1991;17:1152-60.
- 14 Omoto R, Kyo S, Matsumura M, *et al.* Biplane color transoesophageal Doppler echocardiography (color TEE): its advantages and limitations. *Int J Card Imaging* 1989;4:57-8.
- 15 Stümper O, Fraser AG, Ho SY, *et al.* Transoesophageal echocardiography in the longitudinal axis: correlation between anatomy and images and its clinical implications. *Br Heart J* 1990;64:282-8.

Jaundice at 14 days of age: exclude biliary atresia

The majority of infants with biliary atresia in the UK are still being referred too late to get optimal benefit from the operation of portoenterostomy. It is now 14 years since we first reported the importance of early referral in this journal.¹ The success of early surgery has been reported repeatedly since then. Over 80% of infants with biliary atresia who underwent surgery before 60 days of age have become jaundice free, compared with 20-35% for those with later surgery.² Results of surgery are less satisfactory in centres operating on few cases.³ It was clear even 14 years ago that infants who cleared their jaundice had a good prospect of long term survival with a good quality of life.¹ This has been confirmed in the last few years from Japan and Europe.⁴ A 15 year survival of 87% has been reported in infants who become jaundice free.⁵ The age at portoentero-

stomy dictates the frequency of survival beyond 10 years of age. For example in one series 17 of 26 (73%) infants operated by 60 days of age survived to 10 years as compared with only eight of 71 (11%) operated after 90 days of age. Thirty of 48 aged from 10 to 33 years have had a completely uncomplicated course while 37 are leading normal lives with no current medical or surgical problems.⁶ Reports from France are similar with the most satisfactory outcome in those operated on by 45 days of age.⁷ Preliminary analysis of patients treated since 1973 at King's College Hospital suggest a similar outcome but few were treated before 45 days.

These observations on the outcome of early surgery are in keeping with what is known of the pathology of biliary atresia.⁸ In the extrahepatic biliary tract there is a pro-

gressive destructive sclerosing inflammatory process. The intrahepatic bile ducts are also involved leading rapidly to a biliary cirrhosis with a mean age of death of 11 months. Excision of the extrahepatic ducts at a stage when there are still patent biliary channels in the porta hepatitis allows the liver to recover sufficiently for normal growth and development even if the liver is fibrotic and the intrahepatic bile ducts are abnormal. Portal hypertension, present in all cases at the time of surgery, can remit but may still cause alimentary bleeding in 10–15%.⁹ Further liver injury may be caused by cholangitis,¹⁰ but this may be less damaging if it occurs more than five years after surgery.¹¹

For the child who does not have a successful portoenterostomy liver transplantation is the other mode of management. The results of liver transplantation are steadily improving with a one year survival rate after one or more transplantation procedures of over 80%.¹² A five year survival of more than 60% has been reported in older children but less than 40% in infants.^{13 14}

Liver transplantation requires many more resources than portoenterostomy. The patient requires lifelong immunosuppression with close supervision for both medical and surgical complication and the longer term prognosis is unknown. Although an important mode of management for end stage liver disease and metabolic disorders, the role of liver transplantation in biliary atresia is secondary to that of portoenterostomy.¹⁵

Despite several publications in this journal, and elsewhere, we find no evidence that infants born with biliary atresia in the UK are being referred earlier. In 1989 the *Lancet* published the findings of a detailed study of the factors that might influence the outcome of surgery and cause delayed referral.² All 50 UK born infants treated for biliary atresia at King's College Hospital in 1985–7 were analysed. Only the age at surgery predicted the outcome. In the three years covered by the study 81%, 54%, and 73% were referred after 6 weeks of age and 46%, 31%, and 35% after 8 weeks of age. All had been jaundiced with persistently yellow urine from the first week of life. One third of families had been reassured repeatedly by midwives, health visitors, or family doctors that the jaundice was physiological. In these instances referral was often initiated at the well baby review at 6 weeks of age. In 60% of cases inappropriate management by hospital paediatric staff contributed to the delay in referral. For example, no investigations to exclude underlying hepatobiliary disease were performed in seven infants with jaundice after 2 weeks of age or in four with jaundice and vitamin K responsive haemorrhagic diathesis. In five instances a diagnosis of breast milk jaundice was made, although this form of jaundice produces an unconjugated hyperbilirubinaemia with no clinical or biochemical evidence of liver disease. In seven instances the clinician was misled by a fall in the serum bilirubin concentration of up to 40 $\mu\text{mol/l}$. A leading article in the *British Medical Journal*¹⁶ and letters in the correspondence columns of both the *British Medical Journal* and the *Lancet* have stressed the measures necessary for the recognition of suspected cases of biliary atresia by 6 weeks of age.

Of 25 infants born in the UK in 1990 whom we have treated for biliary atresia, 63% were referred after 6 weeks of age and 33% after 8 weeks of age, that is, no younger than the age of referral in 1985–7. Although 70% have had a significant drop in their serum bilirubin concentrations after surgery, as yet it is normal in only 50%. We have no reason to suspect that the reasons for delayed referral in the 1990 cohort are in any way different to those in earlier years. At least two were considered to have breast milk jaundice. Parents are angry when they learn that the outcome of surgery may be jeopardised by the time of referral. It is

embarrassing and possibly imprudent in these days of litigation to delve into reasons for delayed referral. The introduction of new techniques^{17 18} and the twice weekly performance of α_1 -antitrypsin phenotyping have excluded genetic, infective, and endocrine disorders more rapidly, reducing the median time between referral and surgery from 14 to eight days. It is difficult to see how the interval can be reduced further if unnecessary laparotomy, with its risks,¹⁹ is to be avoided.

What is required to ensure earlier referral of infants with biliary atresia in the UK?

(1) *The urine of jaundiced infants must be tested for bilirubin and the direct or conjugated bilirubin measured*

The vast majority of infants with biliary atresia are entirely well during the first four to eight weeks of life apart from mild jaundice. The serious nature of the jaundice will be appreciated only if conjugated hyperbilirubinaemia is found. It is always pathological. It must be suspected in any jaundiced infant in whom the urine is yellow as opposed to colourless or clear. All infants with conjugated hyperbilirubinaemia require early identification to minimise the risk of bleeding from vitamin K malabsorption and so that appropriate investigations can immediately be undertaken to identify disorders for which specific medical or surgical treatment is available. It may be the first indication of genetically determined disorders, the identification of which is of considerable importance for the family.²⁰

(2) *The stool must be seen, by the clinician, to determine whether it is yellow or green in colour*

If consecutive stools over a course of two to three days contain no green or yellow pigment biliary atresia is likely. The infant should be referred to a specialised liver centre with the resources to exclude rapidly other causes of complete cholestasis and the surgical expertise to correct biliary atresia effectively. It should be noted that in a minority of patients with biliary atresia the parents give a clear history of the infant passing green or yellow stools before the development of complete cholestasis.

(3) *Systematic screening for hepatobiliary disorders*

Professional education has clearly not succeeded in getting medical staff to perform the two measures that would optimise the management of biliary atresia. Has the time not come to institute systematic screening for hepatobiliary disorders as is done for other relatively rare conditions such as hyperphenylalaninaemia with an instance of approximately 1:4000 live births or congenital hypothyroidism which has a similar incidence?^{21 22} The estimated incidence of biliary atresia ranges from 1:14 000 to 1:21 000 live births.³ The incidence of hepatobiliary disease in early infancy requiring equally early recognition for optimum treatment of infective, metabolic, or endocrine causes is at least five times higher.²³ We would propose that in all infants who remain jaundiced after 14 days of age the urine should be tested for bilirubin and the direct or conjugated bilirubin measured. This could be initiated by the midwife or family doctor. Physiological jaundice almost invariably clears by 14 days of age except in a small proportion of breast fed infants.²⁴ The number of negative tests would be relatively infrequent.

If bile is present in the urine or the conjugated bilirubin is raised the patient should be referred immediately to a paediatrician. If the stools do not contain green or yellow pigment over the course of two to three days the patient

should be referred to a specialist liver centre able to perform portoenterostomy. This change in management could be facilitated with posters emphasising the importance of prolonged jaundice, yellow urine, and the stool colour as early signs of treatable hepatobiliary disease in antenatal, postnatal, and welfare baby clinics or family practitioner centres in which mothers and babies are seen.

(4) *Changing the age of well baby review from 6 to 4 weeks of age*

Consideration should also be given to changing the age at well baby review from the present 6 weeks to 4 weeks. As well as identifying infants with hepatobiliary disorders it would allow earlier detection of some congenital heart defects, particularly left to right shunts and of subluxation of the hips which may be difficult to detect in the neonatal period. Disturbances in mother-child relationships might be more easily remedied if observed at 4 rather than 6 weeks of age. Since these last two measures have been introduced in Japan the age of infants presenting for surgical correction of biliary atresia has gradually fallen and the outcome has steadily improved.²⁵

Until such measures are instituted in the UK it is essential that paediatricians ensure that the direct bilirubin be measured in all neonates passing through their care, particularly those in whom jaundice is still detectable at 14 days of age. It would not be necessary to screen all well babies, only those who remain jaundiced after 14 days of age. Physiological jaundice almost invariably clears by then except in a small proportion of breast fed infants.²⁴ The urine should be tested for bilirubin and the direct or conjugated bilirubin concentration measured. This could be initiated by the midwife or family doctor.

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- Howard ER, Mowat AP. Extrahepatic biliary atresia. Recent developments in management. *Arch Dis Child* 1977;52:825-7.
- Mieli-Vergani G, Howard ER, Portmann B, Mowat AP. Late referral for biliary atresia—missed opportunities for effective surgery. *Lancet* 1989;ii:421-3.
- McClement J, Howard ER, Mowat AP. Results of surgical treatment for extrahepatic biliary atresia in United Kingdom 1980-82. *BMJ* 1985;290:345-7.
- Anonymous. Long term survival in biliary atresia. *Lancet* 1989;ii:597-8.
- Ohkohchi N, Chiba T, Ohi R, Mori S. Long-term follow-up study of patients with cholangitis after successful Kasai operation in biliary atresia: selection of recipients for liver transplantation. *J Pediatr Gastroenterol Nutr* 1989;9:416-20.
- Ohi R, Nio M, Chiba T, Endo N, Goto M, Ibrahim M. Long-term follow-up after surgery for patients with biliary atresia. *J Pediatr Surg* 1990;25:442-5.
- Gautier F, Laurent J, Bernard O, Valayer J. Improvement of results after Kasai operation. The need for early diagnosis and surgery. *5th international symposium on biliary atresia*. Professional Postgraduate Services, Japan. (in press).
- Howard ER. Biliary atresia. In: Blumgart LH, ed. *Surgery of the liver and biliary tract*. Edinburgh: Churchill Livingstone, 1988:707-22.
- Stringer MD, Howard ER, Mowat AP. Endoscopic sclerotherapy in the management of esophageal varices in 61 children with biliary atresia. *J Pediatr Surg* 1989;24:438-42.
- Houwen RHJ, Zwierstra RP, Severijnen RSVM, et al. Prognosis of extrahepatic biliary atresia. *Arch Dis Child* 1989;64:214-8.
- Gottrand F, Bernard O, Hadchouel M, Pariente D, Gauthier F, Alagille D. Late cholangitis after successful surgical repair of biliary atresia. *Am J Dis Child* 1991;145:213-5.
- Sokal EM, Veyckemans F, de Ville de Goyet J, et al. Liver transplantation in children less than 1 year of age. *J Pediatr* 1990;117:205-10.
- Starzl TE, Demetris AJ, Van Thiel D. Liver transplantation. *N Engl J Med* 1989;321:1014-22.
- Starzl TE, Demetris AJ, Van Thiel D. Liver transplantation. *N Engl J Med* 1989;321:1092-9.
- Alagille D, Laurent J, Roy CC. Is there still a place for the Kasai procedure in the treatment of biliary atresia? *J Pediatr Gastroenterol Nutr* 1989;9:405-6.
- Nelson R. Managing biliary atresia. Referral before 6 weeks is vital. *BMJ* 1989;298:1471.
- El Tumi MA, Clarke MB, Barrett JJ, Mowat AP. Ten minute radiopharmaceutical test in biliary atresia. *Arch Dis Child* 1987;62:180-1.
- Wilkinson ML, Mieli-Vergani G, Ball C, Portmann B, Mowat AP. Endoscopic retrograde cholangiopancreatography in infantile cholestasis. *Arch Dis Child* 1991;66:121-3.
- Markowitz J, Daum F, Kahn EI, et al. Arteriohepatic dysplasia 1. Pitfalls in diagnosis and management. *Hepatology* 1983;3:74-6.
- Mowat AP. Hepatitis and cholestasis in infancy: intrahepatic disorders. *Liver disorders in childhood*. 2nd Ed. London: Butterworths, 37-71.
- Scriver CR, Kaufman S, Woo SLC. The hyperphenylalaninemias. In: Scriver CR, Beaudet AL, Sly WS, Valle D, eds. *The metabolic basis of inherited disorders*. 6th Ed. New York: McGraw-Hill, 1989:495-546.
- Dumont JE, Vassart G, Refetoff S. Thyroid disorders. In: Scriver CR, Beaudet AL, Sly WS, Valle D, eds. *The metabolic basis of inherited disorders*. 6th Ed. New York: McGraw Hill, 1989:1843-80.
- Dick MC, Mowat AP. Hepatitis syndrome in infancy—an epidemiological study with 10 year follow up. *Arch Dis Child* 1985;60:512-6.
- Maisels MJ, Gifford K. Normal serum bilirubin levels in the newborn and the effect of breast-feeding. *Pediatrics* 1986;78:837-43.
- Ohi R, Hanamatsu M, Mochizuki I, Chiba T, Kasai M. Progress in the treatment of biliary atresia. *World J Surg* 1985;9:285-93.