Thoracic foregut duplications

Congenital duplications may arise anywhere in the gastrointestinal tract. Midgut duplications are the most common; foregut duplications (oesophagus, stomach, and the first and second part of the duodenum) account for approximately one third of all duplications. Foregut duplication cysts arise either as posterior mediastinal oesophageal duplications, or as foregut duplications from within the abdominal cavity and present as an abdominal mass, or extend through the diaphragm to lie within the thorax. These cysts often give rise to respiratory symptoms, though their mode of presentation is protean which presents diagnostic difficulties.

Embryology

Foregut duplication cysts may be either proximal or distal. Proximal or mediastinal oesophageal duplications occur early in embryonic life. The notochord, present from the end of the third week, may split and allow endodermal gut to herniate through the gap, resulting in a cyst or fistula ('split notochord syndrome'). The cyst may interfere with the anterior fusion of the vertebral mesoderm; vertebral anomalies are seen in approximately half of these cases. The cyst may rarely extend into the neural canal (neurenteric cyst).

Distal or infradiaphragmatic foregut duplications may present as an abdominal mass or extend into the thoracic cavity through a defect in the diaphragm, and are often tethered to cervical vertebrae. They usually lie in the right hemithorax and may fistulate into the bronchial tree (bronchopulmonary foregut duplication).

Age and sex incidence

Previous series have found a predominance of foregut duplications in girls especially where there is bronchopulmonary involvement. In 20 patients reviewed at our hospital there was an equal sex incidence. The age at diagnosis ranges from birth to the first decade, with most presenting within 18 months of birth. Occasionally, the diagnosis may be made on antenatal ultrasound. Asymptomatic cysts may escape detection until adulthood.

Clinical presentation

Foregut duplications, because of their anatomical position, frequently present with respiratory symptoms; two thirds of patients present with severe respiratory distress which may be present from birth, or symptoms may be insidious with cough, wheeze, or recurrent respiratory infections. Rarely a duplication cyst lined by gastric mucosa may perforate into the bronchial tree, and the patient presents with respiratory distress and haemoptysis. A third of the patients present with gastrointestinal symptoms — for example, vomiting, haematemesis, and an abdominal mass (due to infradiaphragmatic extension). A duplication which extends through a vertebral defect into the neural canal (a neurenteric cyst), may present with signs of spinal cord compression.

Diagnostic imaging

Foregut duplications often present a diagnostic dilemma. The diagnosis of a duplication cyst may occasionally be made on antenatal ultrasound scan; this applies with gastric duplications. A chest radiograph may show a foregut duplication cyst as a cystic area surrounded by opacification. The cyst may be perihilar or subcarinal and overshadowed by the cardiac silhouette and therefore better demonstrated on a lateral chest radiograph. Ultrasonography may be helpful in defining the cystic nature of the duplication — an echogenic inner rim is diagnostic. Some series have reported a high diagnostic yield with barium contrast studies. Other studies have found this investigation less helpful, although useful in confirming the presence of a mass with displacement of normal bowel. Digital subtraction arteriography may define the vasculature of the duplication cyst. Computed tomography is useful in defining the nature of the cyst and its relationship to other structures. The cyst usually has an attenuation similar to that of water, and a rim of calcification may be visible.

A technetium scan will document the presence of ectopic gastric mucosa lining the cyst, and may aid diagnosis. However, the diagnosis may not be made until operation in some instances.

Associated congenital abnormalities

Other congenital abnormalities may occur with foregut duplication cysts. Vertebral anomalies (scoliosis, hemi-vertebrae, and spina bifida) are the most frequent and result from failure of fusion of the vertebral mesoderm. Multiple intestinal duplications occurring in association with foregut duplication cysts are well recognised as is oesophageal atresia. Other associated congenital abnormalities include cardiac anomalies and pericardial defects.

Treatment and outcome

The treatment of foregut duplication cysts is surgical. Surgery may be difficult due to a common vasculature shared between the cyst wall and the adjacent normal bowel. Thoracotomy and/or laparotomy is usually required, although endoscopic removal has been reported. The outcome is usually excellent.

Histopathology of the lining epithelium

Gastric mucosa is the predominant cell type lining foregut duplication cysts. Ciliated epithelium may persist from an embryonic oesophagus. Neural tissue may also be present in the cyst wall. Duplication cysts are often lined by more than one epithelial cell type. Adenocarcinoma has been reported arising in an intrathoracic duplication of foregut origin.

Conclusion

Foregut duplication cysts are infrequent developmental anomalies. Their presence should be considered in a child with unusual respiratory or gastrointestinal symptoms who does not respond to conventional treatment. Complete surgical excision is required to avoid recurrence or later possible malignant transformation in residual tissue and usually results in an excellent outcome.

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Orthopaedic management of cerebral palsy

Like so many other facets of orthopaedic surgery, the orthopaedic management of the child with cerebral palsy has changed significantly in the last 10 to 20 years. As in many other fields, the technology has advanced rapidly, particularly in the development of the sophisticated assessment of gait in the gait laboratory. As usual with such advances, we have a mass of new data that solves some problems and creates others. Gait laboratories are expensive, labour intensive, and time consuming. They are also limited to children who can walk with or without aids within the confines of the laboratory and can cooperate with those running it. They have added significantly to our knowledge but do not provide an easy answer to the often complex and multidisciplinary problems of the child with cerebral palsy. As a result, the orthopaedic surgeon will find himself working within a multidisciplinary team involving the neuropaediatrician, the physiotherapist, orthotist, and several other professionals involved with the care of these complex patients. At present there are few such laboratories in the UK but it is becoming increasingly difficult for an orthopaedic surgeon to provide satisfactory orthopaedic advice in the walking child without one. A simple well lit walkway with video recording equipment capable of individual frame analysis can provide satisfactory observational analysis of a child's gait (M Pearse et al, European Paediatric Orthopaedic Society, Oporto, April 1994).

In a most important article published in Clinical Orthopaedics and Related Research, Rang and Wright asked the question 'What have 30 years of medical progress done for cerebral palsy?'. This is a very thoughtful provoking article and should be prescribed reading for all those involved in the management of cerebral palsy. Rang and Wright suggest that the success of medical treatment could be measured in terms of (i) reduction of the incidence and severity of cerebral palsy; (ii) reduction of the patient's disabilities; (iii) reduction of the burden for the family and for society, both in human and in financial terms. Some progress has been made in all these areas but reviewing the patients attending my clinic from week to week, there is no cause for complacency and some justification for thinking in some ways we seem to be going backwards. Modern neonatal care has reduced the mortality of low birthweight premature infants but the incidence of brain damage leading to cerebral palsy in this group does not seem to have changed significantly.

In my own practice, it is the severe end of the spectrum in cerebral palsy, namely, the child with severe spastic quadriplegia or, to use the term popularised by Professor Bleck, the total body involved child, which has increased most markedly. In the past orthopaedic management, both surgical and orthotic, was rarely considered appropriate for this group. In simple terms, the orthopaedic problems in children with cerebral palsy now break down to those in the ambulant group, for whom gait analysis is becoming more and more important, and the non-ambulant or largely non-ambulant group, in which quality of life as a non-ambulant person is the most important. Nowadays, in order to provide improved seating and mobility in this group, a stable straight spine and stable hips are considered a prerequisite. The incidence of spinal deformity in this group is high. Control of spinal deformity by bracing is difficult. Surgery is often extensive, time consuming, and without significant hazard. The natural history of hip displacement in this group suggests that over 50% will develop progressive displacement and dysplasia and up to 20% frank dislocation with time. Scrutton suggests that those children who do not pull to standing by the age of 3 years are at significant risk and radiographs should be taken of their hips once a year to monitor progressive displacement, which can occur early and be complete by the age of 5. Abduction splintage is widely used, although its precise role and value have not as yet been clearly evaluated. Early soft tissue surgery performed before displacement, measured by the Reimers migration percentage as greater than 50%, has been shown to be reasonably successful (K R Wood, J A Fixsen, British Society for Children's Orthopaedic Surgery, 1989).

Soft tissue surgery alone, once there is more than 50% displacement and/or significant acetabular dysplasia, is unlikely to be successful and requires major bony reconstruction of the hip, usually in the form of open reduction, femoral osteotomy, and acetabuloplasty. Like major spinal surgery, this is not without its hazards and is a significant ordeal for the patient and the parents. The alternative of...