CDH Study Group
by: Kevin Lally, MD

Repair of congenital diaphragmatic hernia (CDH) has changed from an emergent to a delayed procedure in the last decade. Lack of a large multi-center database has hampered progress in the management of CDH making determination of current standard difficult. The CDH Study Group was formed in 1995 to collect data from multiple institutions in North America, Europe and Australia. Participating centers completed a registry form on all live-born infants with CDH during 1995 and 1996. Demographic information, data about surgical management and outcome was collected. Sixty-two centers participated, with 461 patients entered. Overall survival was 280 of 442 patients (63%). The defect was left-sided in 78%, right-sided in 21% and bilateral in 1%. Subcostal approach was used in 91% of patients, with pleural drainage in 76%. A patch was used in 51% of the patients, with PTFE being the most commonly used material (81%). Mean operative time was 102 minutes, with an average blood loss of 14 cc (0-500 cc). A majority of patients underwent repair between 6 AM and 6 PM (88%). 19% had surgical repair on ECMO at a mean time of 170 hours into the ECMO course (10 to 593 hours). Mean age at operation in patients not treated with ECMO was 73 hours (1 to 445 hours). The data indicates that prosthetic patching of the defect has become common, that after-hours repair is infrequent and that delayed operative repair has become the preferred approach in most centers. Furthermore, the mean survival of 63% indicates that despite decades of individual effort, the CDH problem is far from solved.

Duodenal Stenosis
Congenital partial obstruction of the duodenum can be either intrinsic (membrane, web or pure) or extrinsic (Ladd’s bands, annular pancreas). A significant group (25-33%) is born with Down’s syndrome. This does not entail a higher risk of early mortality unless associated with cardiac malformations. Other associated conditions are malrotation (midgut volvulus is rare due to absent bowel distension and peristalsis), biliary tract anomalies and Meckel’s diverticulum. The diagnosis is suggested in utero by the double-bubble image on ultrasound. Vomiting is the most frequent presenting symptom. UGIS is diagnostic, showing a dilated stomach and first duodenal portion with scanty passage of contrast material distally. Management varies accordingly to the type of stenosis: Ladd’s bands are lysed. Pure stenosis is opened longitudinally and closed transversely (Heineke-Mickulicz). Membranous stenosis is resected. Successful endoscopic membranectomy of duodenal stenosis has been reported. Duodeno-duodenostomy is the procedure of choice for annular pancreas. Diaphragms can rarely be double. Anastomotic malfunction requiring prolonged intravenous nutrition and hospitalization has prompted development of a
diamond shape larger stoma. Tapering or plication of the dilated duodenum is another effective method of improving disturbed transit. Other complications after surgery are megaduodenum with blind loop syndrome, biliary reflux, cholestatic jaundice, delayed transit and bowel obstruction. Early mortality is associated to prematurity and associated malformations. Long-term follow-up is warranted to identify late problems.

References

Bile Peritonitis
Bile peritonitis has been reported after conservative management of blunt hepatic trauma. The source is a major bile duct or peripheral injury. Diagnosis may be delayed for several days when the child insidiously develops symptoms of abdominal pain, jaundice and fever. CT-Scan shows the liver fracture with abdominal effusion. ERCP can rule out the extrahepatic origin of the problem. Percutaneous drainage with antibiotic has been successful management with peripheral liver injuries. Most cases explored will need cholangiography to identify a major extrahepatic bile duct injury.

References

*******************************************************************************
* Edited by: Humberto L. Lugo-Vicente, MD, FACS, FAAP
P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico 00922-0426.
Tel (787)-786-3495 Fax (787)-720-6103
E-mail: titolugo@coqui.net Internet: http://home.coqui.net/titolugo
PSU University Edition: http://www.upr.clu.edu/psu