THORACIC SURGERY Congenital diaphragmatic hernia

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Several genes share roles in diaphragmatic, pulmonary , cardiac and foregut development; consequently , congenital diaphrag matic hernias are associated with lung hypoplasia, pulmonary hypertension, cardiac defects and gastroesophageal reflux, with half having additional anomalies. The pulmonary hypoplasia due to lung compression from herniated abdominal is partly contents (mechanical aetiology) and partly due to genetic causes FOG2, GATA4). Commonly , there is a left posterolateral or (e.g. Bochdalek defect, less commonly a ventral or Morgagni defect. Vincent Alexander Bochdalek , 1801–1883, Professor of Anatomy , Prague, Czech Republic. Giovanni Battista Morgagni , 1682–1771, Italian anatomist, the father of modern anatomical pathology . John Bruce Beckwith , 1933, American paediatric pathologist. Hans-Rudolf Wiedemann , 1915-2006, German paediatrician. Both Beckwith and Wiedemann reported cases of the syndrome independently in 1963 and 1964. Neonatal abdominal wall defects /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF The diagnosis is usually made antenatally . A prognosis based on an observed-to-expected lung–head ratio (ultrasound), total fetal lung volume (magnetic resonance imaging) and whether or not the fetal liver is in the chest informs counselling. After birth, intubation, muscle relaxation and gentle ventilation aim to maintain pH, and oxygen saturation so avoiding right-to-left shunting. Permissive hypercapnia and . Pulmonary hypertension high-frequency oscillation may help may lead to vascular shunting, hypoxia, hypercapnia and cardiac dysfunction. Cardiac dysfunction, assessed by echo - cardiography , may respond to nitric oxide, prostaglandin /uni00A0 E1, milrinone and inotropes, but severe dysfunction requires extracorporeal life support (ECLS). Repair is o ff ered if circulation stabilises. Unlike in traumatic the pulmonary diaphragmatic hernias, urgently reducing the bowel does not improve gas exchange in a congenital diaphragmatic hernia. The defect can be approached from the abdomen or the chest, either open or minimally invasively . The defect may be small, needing only a few sutures, or larger, needing a conical Silastic or GOR-TEX patch. A hernial sac may be present, which may be removed or plicated.

Gastroschisis Environmental risk factors (young maternal age, drug use) Bowel complications common (atresia, matting, volvulus) Associated anomalies rare Exomphalos Frequent association with aneuploidies and other genetic syndromes (e.g. Beckwith–Wiedemann) Other structural anomalies common (e.g. cardiac malformations) Large defects may contain liver (exomphalos major) and require delayed or staged closure

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