Duodenal Atresia

Alternative Names
Congenital Duodenal Obstruction
Duodenal Stenosis

WHO International Classification of Diseases
Congenital malformations, deformations and chromosomal abnormalities

OMIM Number
223400

Mode of Inheritance
Autosomal Recessive

Description
Duodenal atresia is the most common type of congenital small bowel obstruction. Its incidence is approximately 1 in 10,000 live births. It is a rare congenital digestive disorder in which complete closure of a portion of the lumen within the first part of the duodenum is present. The clinical presentation depends on the degree of atresia that is present. It may be suspected by the presence of a "double bubble" due to a dilated fluid-filled stomach and proximal duodenum. This appearance corresponds to the gas-filled "double bubble" seen on postpartum radiographs. A classic sign is bilious vomiting without abdominal distention. High-grade lesions will present within the first few hours or days of life whereas less severe obstructions may permit a child to go months or years prior to diagnosis. Weight loss, dehydration and hypochloremic metabolic acidosis are common.

Duodenal atresia is associated with other congenital abnormalities approximately fifty percent of the time. These include malrotation, congenital heart abnormalities, esophageal atresia, and anorectal and renal anomalies.

Molecular Genetics
In many cases, the etiology of duodenal atresia is unknown. Most cases are sporadic, but a genetic component is suggested by the report of familial cases of polyduodenal atresia with an autosomal recessive pattern of inheritances.

Epidemiology in the Arab World

Lebanon
Mishalany et al. (1970) described 2 children with duodenal atresia, all 4 parents of whom were descendants from one couple related as first cousins. Mishalany et al. (1971) then reported that a third affected child had been born in these kindred. Der Kaloustian et al. (1974) reported yet another affected child.

United Arab Emirates
Al Talabani et al. (1998) studied the pattern of major congenital malformations in 24,233 consecutive live and stillbirth at Corniche hospital, which is the only maternity hospital in Abu Dhabi, between January 1992 and January 1995. A total of 401 babies (16.6/1,000), including 289 Arabs, were seen with major malformation. Sporadic conditions accounted for 26% of the cases. In their study, Al Talabani et al. (1998) observed three patients with duodenal atresia or stenosis in families from the United Arab Emirates. Recurrence was not reported in the families. Al Talabani et al. (1998) concluded that their study was very close to representing the true incidence of congenital abnormalities in the whole United Arab Emirates, as they investigated over 98% of deliveries in Abu Dhabi, the capital of United Arab Emirates.

Nawaz et al. (1998) reviewed the experience in the United Arab Emirates in the management of esophageal atresia (EA) and tracheoesophageal fistula (TEF). In their review, Nawaz et al. (1998) studied 41 patients with EA and/or TEF. Approximately 51% of the patients had associated congenital malformations such as: duodenal atresia, hydrocephalus, trisomy 18,
choanal atresia, anorectal agenesis, and others. In 1999, Nawaz et al. conducted a retrospective study to evaluate patients with neonatal intestinal atresia. Twenty-one consecutive newborns with intestinal atresia were observed between 1982 and 1997. There were 10 patients with duodenal obstruction. None of them had Down’s syndrome, but three had associated anomalies. All of them survived, but postoperatively seven of them required total parenteral nutrition. Nawaz et al. (1999) indicated that seven of their patients with duodenal obstruction were dehydrated at the time of referral. This emphasizes the fact that delay in referral of newborns with neonatal intestinal obstruction is associated with increased morbidity and mortality.

References

Contributors
Ghazi O. Tadmouri: 27.2.2005