



Annular Pancreas

WHO International Classification of Diseases

Congenital malformations, deformations and chromosomal abnormalities

OMIM Number

167750

Mode of Inheritance

Autosomal dominant

Description

Annular pancreas is one of the most frequent congenital pancreatic anomalies formed by a thin band of normal pancreatic tissue that completely or partially encircles the second portion of the duodenum. During embryonic development, the ventral pancreatic primordium moves to the right and rotates posteriorly until it comes to lie to the left of the duodenum, subsequently fusing to the dorsal primordium. Annular pancreas is believed to be due to a defect in the normal rotation of the ventral primordium. In more than two thirds of affected infants there are associated malformations, such as: Down syndrome, intestinal malrotation, duodenal and esophageal atresia, tracheoesophageal fistula, imperforate anus, and a variety of cardiac defects.

Molecular Genetics

Although the genetic and molecular basis for this anomaly remains obscure, Indian hedgehog or sonic hedgehog inactivation in mice has been shown to cause overgrowth of ventral pancreatic tissue, a phenotype strikingly similar to annular pancreas.

Epidemiology in the Arab World

Oman

Weber and Freeman (1999) reported a child with annular pancreas along with Down's syndrome. The child had loss of the third and fourth part of the duodenum and of the proximal

jejunum as well as an apple peel configuration of the remaining small bowel. The complete absence of branches from the superior mesenteric artery impaired the blood supply of the distal duodenum. Presence of the annular pancreas may have impaired the flow through the pancreaticoduodenal arcade, which would normally compensate for the distal vascular occlusion.

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 one case of annular pancreas in a family from the United Arab Emirates. Recurrence was not reported in the family. 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.

References

- Al Talabani J, Shubbar AI, Mustafa KE. Major congenital malformations in United Arab Emirates (UAE): need for genetic counselling. *Ann Hum Genet.* 1998; 62 (Pt 5):411-8.
- Weber DM, Freeman NV. Duodenojejunal atresia with apple peel configuration of the ileum and absent superior mesenteric artery: observations on pathogenesis. *J Pediatr Surg.* 1999; 34(9):1427-9.

Contributors

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