Multiple Intestinal Atresia

Alternative Names
Familial Intestinal Polyatresia Syndrome
FIPA
Hereditary Multiple Intestinal Atresia
HMIA

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

OMIM Number
243150

Mode of Inheritance
Autosomal Recessive

Description
Hereditary multiple intestinal atresia is an extremely rare subgroup of intestinal atresias, with an autosomal recessive mode of inheritance. It presents a unique combination of clinical, radiological and pathological findings and is secondary to a malformative process taking place early in intrauterine life and effecting the whole gastrointestinal tract. Multiple intestinal atresia is mainly characterized by multiple and widespread atresias extending mostly from pylorus to rectum, intraluminal calcifications on plain abdominal roentgenogram, and an invariably fatal outcome. Cystic dilatation of the bile ducts can be present in cases with both complete pyloric and duodenal or proximal jejunal atresia.

Epidemiology in the Arab World

Lebanon
In 1971, Mishalany and Der Kaloustian first reported multiple-level intestinal atresia in two siblings of distantly related parents.

United Arab Emirates
Nawaz et al. (1999) conducted a retrospective study to evaluate patients with neonatal intestinal atresia. Twenty-one consecutive newborns with intestinal atresia were observed between 1982 and 1997. Eleven patients (eight males and three females) had small bowel atresias. Two of them had multiple intestinal atresias and in one of them there was an associated colonic atresia.

References

Contributors
Ghazi O. Tadmouri: 12.12.2005
Ghazi O. Tadmouri: 27.2.2005