

Congenital duodenal stenosis and annular pancreas associated with Down syndrome

Migda M, Migda MS
Civis Vita Medical Center, Toruń, Poland

Objective

Down syndrome is recognised as one of the most common predisposing conditions for congenital heart and gut disease. Gastrointestinal abnormalities are frequent, the most common being duodenal stenosis, pyloric stenosis, gastroesophageal reflux, annular pancreas, imperforate anus and Hirschsprung disease. The annular pancreas is a band or ring of pancreatic tissue around duodenum and usually spans close to Vater's papilla. Ultrasound diagnosis is based on detection of so-called double-bubble sign.

Methods

35 year old patient, multipara, underwent prenatal consultation 11-14 wks. Patient underwent prenatal consultation due to previous pregnancy genetic disorder. CRL-73mm, NT-2, 5mm, NB/+/ , TR-/ / DV PI/1, 26/. Risk T. 21 >1: 49. Patient declined invasive testing. In the second trimester of pregnancy double bubble sign was present, no polyhydramnios. After delivery baby had phenotypic signs of Down Syndrome, what was confirmed in postnatal genetic. The newborn was transported to Pediatric Intensive Care Unit due to duodenal atresia (pict 2.). Patient underwent laparotomy– during surgery annular pancreas and Meckel diverticulum were diagnosed. Duodeno-jejunal bypass and Meckel amputation were performed. On day eight, intestinal perforation was present (pict 3.) - the place of Meckel amputation. Second laparotomy was performed. Patient recovered well and discharged.

Results

Case report.

Conclusion

Ultrasonography is an invaluable tool in diagnosing duodenal atresia. Especially associated with polyhydramnios, which suggests compromise with the swallowing problem. Annular pancreas is a rare and is also associated with spectrum on congenital disease like Down Syndrome. This condition may be completely asymptomatic even in the adulthood. More importantly, 40% of annular pancreas cases are associated with life-threatening duodenal atresia or obstruction. Prenatal diagnosis of annular pancreas is usually done based on well known double-bubble sign.