Abstract:

BACKGROUND: Pancreatic agenesis is a rare cause of neonatal diabetes mellitus and the knowledge about the clinical features is sparse. A patient with pancreatic agenesis and double outlet right ventricle is reported. This association has not previously been reported. In addition a synopsis of the patients (n = 14) with pancreatic agenesis who have hitherto been described is given. METHOD: We studied one patient and obtained information on 13 additional patients with pancreatic agenesis by reviewing literature. RESULTS: Literature review: In one patient the pregnancy was terminated at 19 weeks. 31 % (4/13) of the infants died in the first week and 69 % (9/13) in the first six weeks of life, 17 % (2/12) were born preterm and 83 % (10/12) at term, 93 % (13/14) had severe intrauterine growth restriction, onset of diabetes was in 6 out of 10 infants during the first two days of life, ketonuria is rare and has been reported only once. 64 % (9/14) of the infants with pancreatic agenesis had additional malformations mainly of the biliary system (50 %) and/or the heart (36 %). 31 % (4/13) of the infants survived the neonatal period and developed normally. Failure to thrive was compensated by catch-up growth after replacement of pancreatic enzymes and surgical correction of the cardiac malformation. CONCLUSIONS: Pancreatic agenesis is a clinical entity characterized by severe intrauterine growth retardation, early onset of permanent neonatal diabetes mellitus without ketoacidosis, failure to thrive

Pancreatic agenesis as cause for neonatal diabetes mellitus.
due to pancreatic exocrine dysfunction and associated malformations mainly of the biliary system or of the heart. Because of the high neonatal mortality, awareness of pancreatic agenesis as a possible cause of severe intrauterine growth restriction is important for the optimal treatment of diabetes mellitus, exocrine pancreatic insufficiency and the associated malformations.

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