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Agenesis of pancreas and gall-bladder in an infant of incest

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Fatal diabetic coma was observed in a small-for-gestational-age newborn of brother-sister incest. Post mortem examinations revealed congenital absence of the pancreas and gall-bladder.

A variety of congenital abnormalities has been described in children of incest [1, 2]; the present report deals with agenesis of pancreas and gall-bladder in an infant born to siblings.

Report of a Case

A newborn girl was admitted with severe intrauterine dystrophy. Delivery was near term but the baby's birth weight was only 1750 g. She appeared normal physically and the first three days of her life were uneventful. On the fourth day the blood glucose level was 270 mg/100 ml, and glucosuria was found. Between five and eight days of age, the infant was symptomless, her blood sugar varied from 90 to 288 mg/100 ml. On the seventh day the intravenous glucose tolerance test resulted in a diabetic curve, the utilization constant was $Kg = 0.003$. The cerebrospinal fluid was normal except for a glucose content of 153 mg/100 ml. Serum amylase activity was low, 30 Somogyi units. From the ninth day, because of increasing hyperglycaemia and glucosuria without ketosis, insulin was prescribed. In spite of that, diabetic coma developed with a blood glucose peak of 765 mg/100 ml, and the baby died at the age of 14 days with signs of bronchopneumonia.

At necropsy, absence of the gall-bladder and pancreas was noted. Liver structure and bile ducts were normal. The common sites of heterotopic pancreas [5] were thoroughly investigated histologically, but no pancreatic tissue was detected in the gastrointestinal tract including Meckel's diverticulum, in the mesentery and in the mediastinum. Except for bilateral bronchopneumonia, the other organs proved to be normal.

The mother of the child was a 16-year-old unmarried girl. This was her first pregnancy. The course was normal, no teratologic effects could be detected. She confessed that the baby's father was her own 19-year-old brother. Both parents and all their relatives were healthy; neither diabetes nor infantile death had occurred in the family.

Discussion

When admitted, the baby was thought to have transient neonatal hyperglycaemia. Unfortunately, the

blood insulin level has not been determined.

The presence of heterotopic pancreatic tissue is difficult to exclude, but in this infant no aberrant pancreas could be found by meticulous histological examination. Besides, heterotopic pancreas is often associated with elevated serum amylase levels [3] and hypoglycaemia [4], and in our patient the contrary was observed. Thus, in this case a true agenesis of the pancreas had to be assumed, a condition of extreme rarity.

Absence of the gall-bladder is of little pathological significance [7]; its frequency has been estimated at 0.65 per 1000 [6].

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