IS-76 Phosphoenolpyruvate Carboxikinase (PEPCK) Localizes to Proximal Tubules in Near Term Non Human Primate Fetal Kidney

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[Objective] Gluconeogenic capacity of the kidney has been linked to diabetic renal pathologies. While glucose-6-phosphatase has been localized within human fetal kidney proximal tubule cells, little is known about the localization of PEPCK 1 and 2 during renal development in primates. We evaluated the precise localization of the key gluconeogenic enzyme, PEPCK in the non human primate fetal kidney. [Methods] All procedures were institutionally approved. Fetal kidneys were collected at Cesarean section under isoflurane general anesthesia at 165 days gestations (dG; term = 184dG), fixed in 4% paraformaldehyde, and paraffin sections (5um) stained for PEPCK1 and PEPCK2. [Results] PEPCK1 (cytoplasmic isoform) was expressed in S1 proximal tubules. No staining was observed in the medulla or collecting ducts. PEPCK2 (mitochondrial isoform) was found in proximal tubules with the strongest staining in the S1 region. Expression of both isoforms close to the nephrogenic zone was more abundant than in the inner cortex. [Conclusion] There are several reports describing expression of PEPCK in liver during fetal life. This study shows the precise localization of cytoplasmic and mitochondrial PEPCK in fetal non human primate kidney. The role these enzymes play in renal glucose handling remains to be examined.

IS-77 Meckel-Gruber syndrome: Case report

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Objective: Meckel–Gruber syndrome is an autosomal recessive syndrome. The syndrome is rare and lethal. The incidence is 1:10,000 live births. We report a case in which the diagnosis of Meckel–Gruber syndrome was possible by prenatal sonography. Methods: This 34-year-old female, gravida 2 para 1, with unremarkable medical history was referred at gestational age 14 weeks 3 days from local clinic due to nomaity of fetal head. Ultrasound showed absent calvarium above orbits, large kidneys, polydactyly and clubfoot. The amount of amniotic fluid was normal. The parents decided to terminate the pregnancy. Results: Acrania, cystic dysplasia of the kidneys, clubfoot and polydactyly were noted after delivery. The karyotype was normal, and trisomy 13 was excluded. Conclusion: Meckel Gruber was first described by Meckel in 1822 and later by Gruber. It was a syndrome characterized by posterior encephalocele, polydactyly, and cystic dysplasia of the kidneys. Other abnormalities which can occur in association with the syndrome and which may be detectable sonographically include micrognathia, cardiac abnormalities, syndactyly, clinodactyly, and clubbed foot. Families with previously affected children stand a 25% chance of recurrence in future pregnancies. Prenatal diagnosis has been extremely important for those families. Trisomy 13 which is mostly a sporadic event with a rather low recurrence rate is the most likely syndrome. A karyotype, therefore, is important. The triad of encephalocele, polydactyly, and large echogenic kidneys seen prenatally should suggest the diagnosis of Meckel–Gruber syndrome.

IS-78 Giant Congenital True Pancreatic Cyst: Case Report and Literature Reviews

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Objective: Congenital true pancreatic cyst is very rare. It is difficult to differentiate pancreatic cyst from other abdominal cysts on antenatal exam. Herein we present a case of a fetal pancreatic cyst, detected at 17 weeks' gestation by ultrasound. Methods: A 31-year-old woman, gravida 1 para 0, with unremarkable medical history was referred for evaluation of a male fetus suspected of duodenal atresia. Ultrasound revealed a cystic structure within the right upper abdomen. The cyst was connected with a tubular formation. There was no polyhydramnios. Choledochal cyst was suspected at first. At 17 weeks' gestation, a genetic amniocentesis was performed that revealed a normal 46, XY karyotype. Subsequent ultrasound examination at 22 and 34 weeks' gestation showed the size of cyst was increasing rapidly. A 3,026 gm male neonate was born at 34 weeks' gestation by Cesarean section. Results: Exploded laparotomy was performed at Day 4. One giant cyst was contacted with great gastric curve, omentum, and duodenum. Cystectomy was performed. Pathological report revealed that it was true pancreatic cyst with pancreatic lining. Conclusion: Pancreatic cyst was extremely rare. We should make a differential diagnosis from duodenal atresia, choledochal cyst, liver cyst, omental cyst, and lymphangioma. 3D ultrasound will be helpful to diagnosis.