and 7 girls and CAH in 50% of the boys but none with hypertension.

The child is under treatment with hydralazine and prednisolone. On nine months follow up, the hypertension is under control, the skin pigmentation has disappeared and the linear and genital growth is static.

REFERENCES


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**Choledochal Cyst in a Neonate**

A.K. Basu  
J. Basu  
K. Basu  
S. Banerjee

Choledochal cyst is a relatively rare condition. A review of the literature reveals

From the Departments of Pediatric Surgery, Anesthesiology and Pathology, Medical College, Calcutta.

Reprint requests: Dr. Ashoke Kumar Basu, 90, Ballygunge Place, Calcutta.

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six patients of choledochal cyst who presented in the neonatal period. Four of these six patients were diagnosed antenatally(1,3). We present a child with choledochal cyst in the neonatal period.

**Case Report**

A two-day-old boy weighing 3.9 kg presented at Park Children’s Centre for Treatment and Research, Calcutta with history of distension of abdomen since birth and nonbilious vomiting for last 24 hours. He was passing high colored urine and had passed meconium on the second day of life. On examination he was mildly icteric. Examination of the abdomen revealed a large cystic lump extending from the right hypochondrium to the right iliac fossa and crossing the midline at the level of the umbilicus. It had slight side to side mobility and was dull on percussion. Plain X-ray of the abdomen showed a soft tissue shadow occupying the whole of the right half of the abdomen with intestinat gas shadows pushed to the left (Fig. 1). Ultrasonography showed a cystic lesion occupying the whole of the right half of the abdomen (Fig. 2). Intrahepatic biliary channels were dilated. A diagnosis of choledochal cyst was suggested. His hematological and biochemical parameters were within normal limits for his age except a raised total bilirubin level of 3.7 mg/dl (unconjugated 1.4, conjugated 2.3).

Peroperative preparation included administration of injection vitamin K, intravenous glucose infusion and decompression of the bowel by nasogastric suction and bowel washes. Laparotomy was performed on the 5th day of life. Abdomen was opened through right transverse supraumbilical incision. A unilocular cystic lump measuring 15 cm × 10 cm was seen
occupying the whole of the right half of the abdomen. The cyst had extended behind the duodenum which was stretched and obstructed explaining the early nonbilious vomiting. The liver and the gall bladder looked normal. The cyst could be completely decompressed through cholecystotomy. The cyst contained dark brown fluid. Since it was a large cyst in a small patient, complete excision of the cyst was considered hazardous. A 2.5 cm wide single layer choledochocystoduodenostomy was performed. Biopsies were taken from the gall bladder, liver and cyst wall. A cholecystostomy was performed with a Malecot catheter and abdomen was closed in layers. Post-operative period was uneventful. Contrast study on 7th postoperative day through the cholecystostomy showed free flow of dye into the duodenum with no hold up in the cyst. The Malecot catheter was removed.

Microscopically the gall bladder and the liver were normal. The cyst showed fibrous tissue wall lined with cuboidal epithelium. There were areas of hemorrhage. Ultrasonography repeated two months after surgery showed normal size common bile duct with no evidence of cyst. In one and half years follow-up the patient
had no attack of cholangitis and is thriving normally. His liver function tests are also normal.

Discussion

Earliest diagnosis of choledochal cyst has been made at 15th week of gestation(3). Three more patients have been diagnosed by antenatal routine ultrasonography(4,6). Review of the literature reveal two more cases of neonatal choledochal cyst(1,2). Perhaps ours is the seventh case of neonatal choledochal cyst. Many different mechanisms for development of choledochal cyst have been suggested. These include (i) Failure of canalization of primordial biliary tree(7), (ii) Localized stenosis secondary to focal mural abnormality or external pressure or due to anatomical kinking(6), (iii) Damage to the ducts at an early gestational age due to reflux of activated proteolytic pancreatic enzymes from a common extrudodental channel into the lower end of the common bile duct(6). Ultrasonography and radionuclease scanning are the two most rewarding investigations for preoperative diagnosis of choledochal cyst(8). In our case ultrasonography was most helpful. Routine preoperative cholangiography can demonstrate the presence of obstruction or the presence of anomalous pancreaticobiliary communication. We did not perform any intraoperative contrast study in order to cut short the operating time. The accepted treatment for choledochal cyst is excision of the cyst and Roux-en-Y Hepaticocholedochojunostomy(1,9). This was not possible in our patient because the patient was only five days old and the cyst was 15 x 10 cm in size. Extensive dissection in such a patient could prove to be counterproductive. The procedure of internal drainage of the cyst leaves behind the cyst and hence there may be attacks of cholangitis or carcinoma in future. We are prepared to do a revision surgery of cystectomy and hepaticocholedochojejunostomy in our patient if he gets attacks of cholangitis. However, in a follow-up of one and half years there has been no episode of cholangitis. A recent report from Japan(10) has suggested that metaplastic changes occur in the cyst epithelium secondary to cholangitis and repeated destruction and regrowth of the lining epithelium. Completion of neoplastic transformation requires decades of exposure to this process of destruction and regrowth. Since we are prepared to do cyst excision in our patient if he gets cholangitis the chance of carcinoma developing in our patient is remote.

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Renal Glycosuria in Two Siblings

P.V. Havaladkar
N.S. Mahantshetty
B.M. Siddibhavi

Renal glycosuria denotes the renal tubular abnormality in individuals who excrete a variable amount of glucose in the urine at normal levels of blood glucose. The age at which the disorder is first detected varies, but in the majority of patients it is in the second decade(1). Though in one instance it was discovered in a 4 weeks old patient(2), reporting in pediatric age is less frequent. Even with the use of titration method for renal filtered glucose, different observers reached opposite conclusions about the possible pattern of inheritance, both autosomal dominant and recessive trait being interpreted(1). Two siblings diagnosed as renal glycosuria as per the criteria laid by Marble and Ferguson(3) are described.

Case Report

A 3-year-boy was admitted with complaints of abdominal pain of 15 days duration. There was no history of polyuria, polyphagia and polydipsia or dysuria. He was anemic and systemic examination did not show any positive findings. His blood counts were within normal limits. Urine was persistently positive for reducing substance in all the samples taken in a day. It was confirmed to be glucose by dipstick method and osazone preparation. Blood sugar after overnight fast was 83 mg/dl and GTT was normal. Blood urea was 25 mg/dl and serum creatinine 0.8 mg/dl. Urine culture did not grow any organisms and ultrasonography of kidney was normal. Urinary excretion of bicarbonates, phosphates and aminoacids were within normal limits and also the acid-base studies in the blood. Urine sugar was persistently positive on follow up of 2 years. A 13-year-old sister and 10-year-old brother were screened and found to be normal. However, a 7-year-old sister had asymptomatic persistent glycosuria with normal fasting blood sugar and GTT. Other investigations were also normal. They were born to non-consanguineous parents. Both parents when screened for glycosuria were negative. There was no family history of Diabetes mellitus.

Discussion

The criteria(3) used for the diagnosis of renal glycosuria are (i) constant glycosuria