of high fever in warm environment, dry skin, sparse hypopigmented hair, frontal bossing, flattened nasal bridge are some of other features. Poor development of mucous glands in the respiratory tract may result in increased susceptibility to respiratory tract infections. Treatment of these children include protecting them from high ambient temperatures and early dental evaluation so that prostheses can be provided. Artificial tears should be used to prevent the damage to cornea. Wigs may be advised to improve appearance. (1) Both the brothers reported here had the classical triad along with other clinical features of hypohidrotic (anhidrotic) ectodermal dysplasia which was confirmed by skin biopsy. The mode of inheritance of hypohidrotic ectodermal dysplasia is X-linked recessive with full expression only in males; however, an autosomal recessive mode of inheritance may be operative in some families (1,5).

REFERENCES
2. Smagrupps Centrum - the Swedish Information Center for Rare Diseases- produced and edited this information material. Date of publication: 2005-05-03 Version: 2.1 Article number: 2002-110-10

LETTER TO EDITOR (VIEWERS CHOICE)
VENTRICULOPERITONEAL SHUNT CATHETER MIGRATION THROUGH UMBILICUS - A RARE COMPLICATION

Dipankar Sarkar, Shruti Sarkar

Figure 1: Distal end of VP shunt protruding from the umbilicus

A 9 month old baby was brought to our outpatient department with the complaint of a plastic tube protruding out of abdomen through umbilicus for a few days. The baby had undergone Venticuloperitoneal shunt surgery nearly 4 months back for congenital hydrocephalus in another hospital. He was born at term by caesarean section and was diagnosed to have congenital hydrocephalus soon after birth. The baby had recovered well from surgery, was feeding well on demand and thriving. On examination the baby looked well and the VP shunt catheter was seen protruding out of umbilicus (Figure 1). On pressing the shunt reservoir behind the ear, fluid started dribbling out of the tube protruding through the umbilicus confirming the protruding end to be the distal end of the VP shunt. Systemic examination apart from this did not reveal any other abnormality. The infant along with his parents had come from a distant village. The parents were explained in detail the occurrence of this complication and the need for correction by surgery. They declined corrective surgery and were lost to subsequent follow up.

Migration or protrusion of the ventriculoperitoneal shunt catheter to various sites is not very uncommon and has been reported in several case reports. Cases of shunt migration into stomach (1), bowel (2), liver (3), chest (4), jugular vein (5), anus (6), vagina (7), and scrotum (8) have been reported. In 1973 Adeloye et al reported a 9 month old baby with shunt migration through umbilicus within 2 months after surgery (9). Another case has been reported from Srinagar, India in 2000 where an 18 month old child presented with low grade fever, irritability, abdominal pain along with the distal end of VP shunt coming out through the umbilicus (10) .This particular complication can be explained on anatomical basis. Umbilicus is a centrally situated scar
and it becomes an area of anatomical weakness when there is a foreign body inside the abdomen. There are five ridges in the peritoneum that line the lower part of the anterior abdominal wall, which converge in the midline at the umbilical region. Peristaltic activities can thus direct foreign bodies in the peritoneal cavity toward the umbilicus and push it to come out (9). Migration of the lower end of the shunt catheter is an infrequent problem, which occurs without any recognizable cause. The reported incidence of distal shunt migration is 10% (11). Of all the sites of distal end migration reported so far umbilicus is very rare.

References

From: Department of Pediatrics, Peoples College of Medical Science & Research Center, Bhopal, MP
Address for Correspondence: Dr Dipankar Sarkar, HIG-13, A-Block, Peoples Medical College Campus, Bypass Road, Bhanpur, Bhopal, MP-462010. Email: dipankarshruti@gmail.com
E-published: March 2010

LETTER TO EDITOR (VIEWERS CHOICE)

CONGENITAL ADRENAL HYPERPLASIA IN A MALE CHILD

Manju Mittal, Kumar Ankur, Praveen Kumar, B. Rath

An one and half month old male baby was brought in cardio respiratory arrest with a history of noisy breathing for 3 days and several episodes of vomiting and watery loose motions for 1 day. Baby was immediately resuscitated. His birth weight was 2.5 kg and perinatal course was uneventful. There was one previous hospitalization at day 21 of life with acute gastroenteritis and sepsis. Physical examination revealed normal hemogram, genitalia. Investigations revealed normal hemogram, hypoglycemia (Blood sugar = 24mg/dl), positive CRP, hyponatremia (serum sodium of 118 mEq/dl) and hyperkalemia (Serum potassium = 6.5 mEq/L). These investigations lead us to suspicion of Congenital Adrenal hyperplasia (CAH). Steroid levels were done which showed Serum Cortisol to be low (25.13, normal range = 3-25 ng/dl) and Serum 17 Hydroxy Progesterone = 28-662 nmol/l) and Serum 17 Hydroxy Progesterone (normal range = 100 ng/dl) thus confirming the diagnosis of 21 hydroxylase deficiency. The child was managed with antibiotics, Fludrocortisone and hydrocortisone. Child improved, gained a weight of 500 grams over 10 days of hospital stay. Serum electrolytes normalized (Serum sodium of 140mEq/L and serum potassium of 5.5mEq/L) and child was discharged. At 3 months of follow-up he weighs 3.2 kg and electrolytes were normal.

Congenital adrenal hyperplasia is a group of autosomal recessive disorders resulting from the deficiency of one of the five enzymes required for the synthesis of cortisol (1). This disease is easily clinched in female newborns where the ambiguous genitalia are present, but the diagnosis in male newborns is often overlooked as was in our case. He was admitted at 21 days of life and treated as acute gastroenteritis with sepsis and diagnosis of CAH was missed. There are three types of CAH due to 21-hydroxylase deficiency: classical salt wasting disease which is most severe, both cortisol and aldosterone are deficient, classical simple virilizing disease in which adequate levels of aldosterone are synthesized but adrenal androgens are elevated and non-classical disease in which adrenal androgens are mildly elevated leading to signs of androgen excess after birth. Progressive weight loss, anorexia, vomiting,