fundus of the bladder into the peritoneal cavity (Fig. 2). Thereafter urine continued to drain freely from the catheter, and the child was successfully weaned off the ventilator. Ascites accumulated again and on the second day of life a further 12-hour period of assisted ventilation was required. On the fifth day the infant was transferred to this hospital for further assessment.

Repeat abdominal paracentesis was performed and thereafter the abdominal distension gradually lessened and urine drained freely in the urethral catheter. No surgical intervention was thought necessary at that time. Recovery was temporarily slowed by three complications—paralytic ileus, transient renal tubular defect with polyuria and urinary sodium loss, and coliform urinary infection. Each responded satisfactorily to appropriate therapy. The urethral catheter was removed on the 13th day, and she was able to pass urine normally thereafter.

The child is now 7 months old, is well, and has normal renal function. A repeat intravenous urogram has shown completely normal appearances.

Discussion

Most cases of urinary ascites may be grouped into three categories (Weller and Miller, 1973). Firstly, there are the majority of patients who have posterior urethral valves and obstruction. A second group of those with complex caudal anomalies, usually anorectal and urethral atresia. A third group of those with miscellaneous lesions which include bladder outlet flaps, ureteroceles, ureteral atresia, myelomeningocele with neurogenic bladder, and extrinsic lesions such as presacral neuroblastoma. Spontaneous rupture of the bladder in the newborn must be extremely rare and our search of published reports showed only 4 reported cases to date (Miller *et al.*, 1960; Gandhi, 1964). Only 2 liveborn females have been described previously with urinary ascites, and both these children had bladder outflow obstruction (Baghdassarian, Koehler, and Schultze, 1961; Howat, 1971).

Bladder rupture in our 2 cases must have occurred *in utero* or possibly during the birth process, though it is difficult to accept that during this premature labour, there would be sufficiently increased hydrostatic pressure within the urinary tract to cause the bladder to rupture. In the second child it is not likely that the uneventful laparotomy can be implicated as a cause of the bladder rupture. A question must be raised as to whether or not there was an inherent weakness of the bladder wall present in these children.

The overall mortality in the few reported series of neonatal ascites has been high. Infants with massive ascites at birth do less well, only 1 of 8 infants in one series surviving, whereas the prog-

nosis is better if ascites develops in the first few days of extrauterine life (Cywes, Wynne, and Louw, 1968).

Summary

Two premature infants who presented with neonatal ascites due to apparent spontaneous rupture of the bladder are described. In both children the site of bladder rupture was clearly shown, but neither at time of presentation nor at subsequent review at age 7 years and 7 months, respectively, was any other renal tract abnormality detected.

We thank Dr. M. M. Kerr, Glasgow, and Dr. S. Wilson, Dundee, for permission to publish these cases, and our surgical colleagues, Mr. J. F. R. Bentley and Mr. D. G. Young for their help.

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Congenital midline scalp and skull defect

Congenital midline scalp and skull defects have been reported periodically since 1826 (Greig, 1931) and the published reports have appeared mainly in America. In this communication we record the presence of this congenital anomaly which has occurred in a family over two generations.

Case report

In the first stage of labour, in this mother's third pregnancy at term, the midwife reported that the fetal skull felt abnormal per vaginam and at delivery two abnormal areas were found to be present across the midline at the vertex. The abnormal areas measured

FIG. 1.—The two defects in the baby's scalp shortly after birth are shown.

1.5 × 2.5 cm and 2.5 × 2.25 cm and these were covered by a thin membrane through which the sagittal sinus was seen to distend with blood when the baby cried (Fig. 1). The remainder of the skull appeared to be normal and there was no evidence of any other congenital anomaly.

In a few days the membrane became opaque and thickened. Crusting occurred within a fortnight and by the age of 5 weeks the areas were covered by pink shiny skin which was hair-free. The palpable bone defects appeared to be much smaller and the baby was well, though slow to feed. Skull x-rays at the age of 6 weeks showed two areas of diminished bone density in the region of the skin lesions.

Within a few minutes of birth the mother was shown her baby and was very unconcerned by the appearance of the scalp defects, saying that she had suffered the same problem at birth. Review of her neonatal notes revealed that she had had two similarly sited lesions. The skin edge was described as being continuous with a transparent membrane through which brain could be seen. Within 9 days her lesions were smaller and crusted, and within 9 weeks the lesions had fully epithelialized with non-hair bearing skin. At age 5 months the skull bones felt normal. Now, 26 years later there are two hypertrophied hairless patches at the vertex (Fig. 2). The mother is otherwise physically well but at the age of 10 years had an intelligence quotient of only 62. There is no history of anyone else in the family suffering from such skull defects over three generations.

Discussion

The first recorded instance of these defects occurring in families was reported in 1826. Since then familial cases have been reported in father and son (Johnsonbaugh, Light, and Sutherland, 1965), in mother and 2 daughters (Frank and Ruby, 1957), and in twin sisters (Hodgman, Mathies,

FIG. 2.—The two hypertrophied hairless areas on mother's scalp aged 26 years are shown.

and Levan, 1965); one of the latter having three lesions and the other one lesion. The defects are usually parieto-occipital and most commonly affect females. Skull x-rays have sometimes shown underlying lytic bone lesions which have closed before the age of one year.

The defects are usually attributed to congenital deficiencies in the process of embryological development. In 1965 a child was reported with defects of midline fusion, and deletion of the short arm of chromosome 4-5 was found (Hirschhorn, Cooper, and Firschein, 1965). At birth a midline scalp skin defect was present and healed within 5 months. There was a midline cleft of the soft palate along with other abnormalities. Absence of the nasal septum was recorded with midline scalp defects in 1931 (Greig). Others have reported the association of such scalp defects with trisomy D (Kosnik and Sayers, 1975). Our patient and her mother both had normal chromosomes. It seems likely then that these lesions have a genetic cause rather than being due to local pressure or trauma.

These skull defects are potentially very dangerous as infection may occur and lead to meningitis, or there may be ulceration and haemorrhage. In at least 2 cases fatal haemorrhage from the sagittal sinus occurred early (Savage, 1956; Greig, 1931). Some centres cover the lesions in tulle gras to give some protection over the first few days of life (Savage, 1956). When first seen the small lesions

tend to look alarming but surgical measures do not seem to be required unless penetration of the sagittal sinus occurs. However, lesions are occasionally very large and measure up to 10 cm in length from anterior to posterior fontanelle. As the extensive eschar on these separates during the first month of life there is more of a risk of haemorrhage, and very early excision of the membrane and covering by rotation of skin flaps has been suggested as the treatment of choice (Lynch and Kahn, 1970). Good growth of hair has been reported after this procedure.

Summary

A family with multiple congenital scalp defects, over two generations and probably genetically determined, is described. Although alarming in appearance, surgical intervention is not indicated at least for small lesions. The risk of haemorrhage and meningitis is emphasized.

We thank Dr. Gerald Neligan, University of Newcastle upon Tyne, for referring the case.

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Reye's syndrome associated with acute tubular necrosis

We report the cases of 2 children with Reye's syndrome (acute encephalopathy and fatty degeneration of viscera), both of whom developed

acute renal failure, a hitherto unreported complication, and in one of whom the syndrome followed an influenza A virus infection.

Case reports

Case 1. A boy aged 6·25 years, became ill with anorexia, diarrhoea, and vomiting. 3 days later he had a grand mal convulsion and was admitted to hospital. His past history included the following. (a) Neonatal convulsions thought to be secondary to a cerebral haemorrhage; he had been left with a left hemiparesis, but his intellectual progress had been normal. (b) Two grand mal convulsions at the age of 3 years, since when he has been taking phenobarbitone.

On admission to hospital he was afebrile and had further convulsions, between which he did not regain consciousness. A lumbar puncture showed clear, cell-free cerebrospinal fluid (CSF) under normal pressure, with a protein content of 15 mg/dl. Blood glucose and calcium were normal. There was no clinical evidence of infection. He was later transferred to The Hospital for Sick Children, London. On arrival he was deeply comatose, with generally increased muscle tone and signs of his pre-existing hemiplegia. The liver was palpable 1 cm below the costal margin. Blood pressure was normal. Haemoglobin was 13·8 g/dl, white cell count 6800/mm³, blood urea 136 mg/dl, potassium 5·4 mEq, sodium 131 mEq, and chloride 99 mEq/l. The platelet count was 83 000/mm³, prothrombin time 39 s (control 14), thrombin time 12·5 s (control 9). Fibrin degradation products were not detected in the blood. Repeat CSF analysis was normal apart from a protein content of 90 mg/dl. The blood lead level was normal; calcium 8·9 mg/dl, bilirubin 2·4 mg/dl, serum aspartate aminotransferase 4300 IU/l, serum alanine aminotransferase 4075 IU/l, blood ammonia 90 µg/dl, and a C3 level of 20% of the standard reference serum. Viral studies showed no evidence of a recent infection with herpes simplex, mumps, measles, or influenza A or B viruses. An electroencephalogram showed severe generalized abnormalities with a gross excess of slow-wave activity.

He initially passed small quantities of urine containing blood and protein, but within 68 hours of admission he developed frusemide-resistant anuria. Peritoneal dialysis was instituted. A renal biopsy performed 2 weeks later showed acute tubular necrosis with tubular regeneration. Renal function recovered spontaneously after 3 weeks' dialysis, and liver function tests reverted to normal. He has remained in coma, with frequent convulsions, and the signs of a spastic quadriplegia.

Case 2. A boy aged 10·25 years, became ill with generalized abdominal pain, nonproductive cough, and pyrexia. 24 hours later he had a prolonged generalized convulsion and was admitted to hospital. There was evidence of influenza A in the local community at this time. His history included (a) hypothyroidism diagnosed at the age of 9 months, since when he had received replacement thyroxine, (b) convulsions with fever at