Prune Belly Syndrome

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SUMMARY

Two cases of prune belly syndrome in Black infants are presented. The prune belly syndrome, or congenital absence of abdominal muscles, is accompanied by hydro-ureter, hydronephrosis, megalocystis and usually undescended testes. Other associated congenital defects occur, of which orthopaedic defects appear to be the most prevalent. Others are patent urachus, congenital heart malformation, anomalies of the eyes and ears, ectodermal dysplasia, torticollis and micrognathia.

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CASE 1

A male Black child presented at 18 days of age with a history of having been born with an abnormal abdomen. Obstetric and family history was normal.

On examination the abdomen was lax and floppy, with lateral distension. Overlying skin was wrinkled and slightly hyperpigmented. Both testes were undescended. No orthopaedic abnormalities were present. The rest of the examination, special investigations and X-ray films of chest, skull and skeleton were normal.

The child contracted gastro-enteritis and died at the age of 46 days.

Autopsy Findings

There were no apparent orthopaedic defects. The skin over the abdomen was wrinkled and hyperpigmented. The musculature of the anterior abdominal wall was virtually absent, with only a thin streak of muscle

visible in the region of the left rectus abdominis. All other abdominal muscles appeared fibrosed.

The heart was small with a patent ductus arteriosus present.

In the peritoneal cavity a marked malrotation of the entire midgut was found. A well-formed mesentery extended from the pylorus along the duodenum to the mid-transverse colon. The caecum was in the upper abdominal cavity attached by a long mesentery. The pancreas also had a mesentery. The stomach was correctly rotated and bound down.

The left kidney was smaller than the right. The bladder —a large linear organ—extended up to the umbilicus. The lower one-third of both ureters was grossly dilated. Both testes were totally undescended, situated as minute organs on the ureters. A Müllerian remnant was clearly demonstrable on the bladder.

The rest of the findings were normal.

CASE 2

A 21-day-old male Black child presented with a history of coughing and cyanosis for 2 days, and an abnormal abdomen since birth. Further history was normal.

On examination the child was pyrexial (37,5°C), and centrally cyanosed without clubbing, jaundice or pallor. Chest examination showed tachypnoea, intercostal recession and signs of consolidation in the left upper lobe of the lung. The abdomen was wrinkled with lateral distension (Fig. 1). Both testes were undescended.

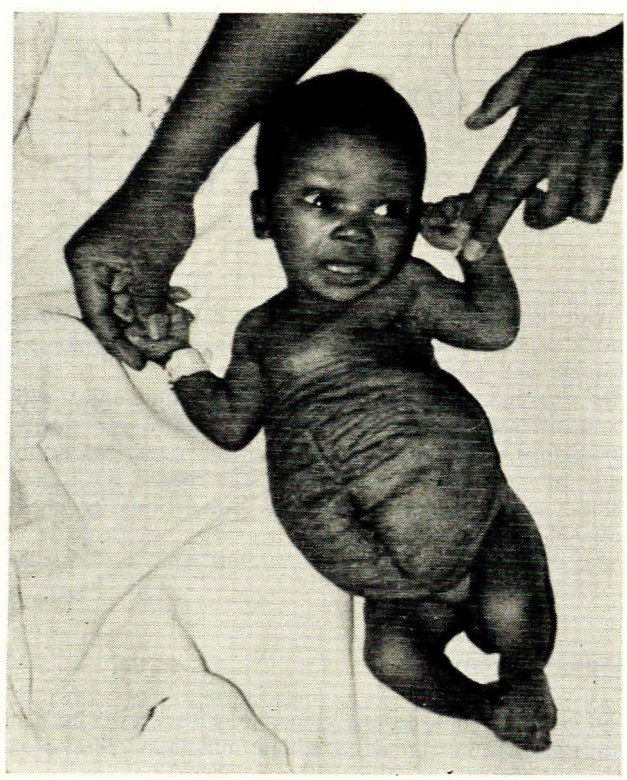

Fig. 1. Case 2—showing 'prune belly'.

Special investigations produced the following abnormal results; urine yielded a pure growth of *Escherichia coli;* a chest radiograph showed consolidation of the left upper lobe and an intravenous pyelogram showed bilateral hydronephrosis.

DISCUSSION

The most obvious anomaly in the condition known as prune belly syndrome is the defect in abdominal musculature with an absence of the lower part of the rectus abdominis and the lower medial parts of the oblique muscles. It was first noted by Fröhlich in 1839.[1] The protruding, thin-walled abdomen and wrinkled skin give the syndrome its name. The chief importance of the condition lies in the other features of the syndrome, first described by R. W. Parker in 1895.[2]

The bladder and ureters are usually greatly dilated with hydronephrosis and small dysplastic kidneys and undescended testes. The anomalies of the urinary tract were first thought to be due to urethral obstruction, but this was present, together with a patent urachus, in only 3 of a series of 20 cases reported.[3] Further, the enlarged bladder does not show the trabeculation usually found with urethral obstruction, and there is probably a primary defect in the musculature of both bladder and ureters. Renal dysplasia is of a type suggesting embryonic dysplasia rather than the effects of back pressure.[4]

Other abnormalities that may be present are talipes equinovarus, congenital dysplasia of the hip, congenital heart malformation, malrotation of the gut, anomalies of the eyes and ears, ectodermal dysplasia, micrognathia and torticollis.

The aetiology is still obscure. The condition is almost entirely limited to males. Of 191 cases reported only 9 were female and they do not appear to have had the same condition.[5] No familial studies are reported.

The prognosis depends largely on the renal function. A follow-up of 14 patients attending the Mayo Clinic since 1945 disclosed 5 patients who had passed the age of 20 years.[6] Of these, 3 were still alive, while 2 had died at the ages of 28 and 24 years.

Surgical procedures including cystostomy, transurethral resection, ileal conduits, nephrectomy and ureteroneocystectomy have been performed, but the survival gain is not significant.[7]

There is little indication that surgery alters the course, provided evidence of obstruction is not present. It is difficult to correlate survival with any single factor or group of factors. A most impressive clinical finding was the persistence of bacteriuria and azotaemia.

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