UNUSUAL CONSTELLATION OF ANOMALIES

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BACKGROUND AND AIMS:

This report details the very rare association of Prune Belly and Pouch Colon in a 34-week twin boy.

CASE PRESENTATION

Baby was born in a level 2 NICU by emergency C-section for maternal abdominal pain. Antenatal scans were suggestive of megacystis and oligohydramnios. Baby was born in poor condition, intubated briefly for respiratory distress syndrome. He was found to have Prune Belly, imperforate anus and bilateral impalpable testes. He was transferred to the regional unit for surgical management. Urethral catheterization allowed for good urine output but his abdomen was distended. Abdominal X-ray showed a large left-sided (partially calcified) mass causing midline shift.

Laparotomy on day two confirmed Type 2 Pouch Colon, where the entire colon is pouch-shaped, ending in a fistula with the bladder. Atretic appendix, Meckel's diverticulum and intra-abdominal testes were also seen. Pouchostomy was successfully formed, enabling early feed commencement. Cystoscopy showed a large, smooth bladder with normal urethra. Intra-operative cystogram demonstrated Grade 3/4 left-sided reflux and tortuous ureter.

Baby recovered well, with plans for distal loopogram and reconstructive surgery at 6 months. Microarray and ultrasound spine were normal.

DISCUSSION

Prune Belly is a spectrum characterised by cryptorchidism, abdominal wall defects and genitourinary abnormalities, occasionally associated with gastrointestinal, cardiac and orthopaedic abnormalities. Pouch colon is a rare anomaly associated with anorectal malformations, seen most commonly in Northern India. The occurrence of both anomalies together is extremely rare, with only a small number of cases previously reported. Enhanced awareness of both conditions will improve detection and early surgical intervention.

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BRILLIANTLY TRANSILLUMINANT ABDOMEN

FIRST X-RAY DEPICTING LARGE ABDOMINAL MASS

DISTAL LOOPOGRAM SHOWING FISTULA INTO THE BLADDER PRIOR TO RECONSTRUCTION

INTRAOPERATIVE VIEW OF POUCH COLON

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