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Bilateral congenital perineal hernias: Spontaneous resolution

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ABSTRACT

A congenital perineal hernia develops through a defect in the pelvic floor muscles and presents as a reducible buttock swelling. These hernias are exceptionally rare with only six cases previously documented in the English literature. We report an extremely premature infant who developed bilateral posterior perineal hernias at 2 weeks of age. The diagnosis was confirmed by ultrasound scan. She remained asymptomatic. By a corrected age of 10 weeks post term, the hernias had resolved clinically and on imaging. A congenital perineal hernia should be considered in the differential diagnosis of a reducible buttock swelling. The diagnosis can be confirmed by imaging and elective surgical repair is indicated for symptomatic or persistent hernias. This case demonstrates that an asymptomatic perineal hernia in a very premature infant should initially be managed conservatively as it may resolve spontaneously.

1. Introduction

Congenital perineal hernias are very rare. Only six cases have been reported before [1–5]. Bowel herniates through a congenital defect in the levator ani muscle of the pelvic floor causing a reducible buttock swelling. Strangulation has not been reported in children, perhaps because the neck of the hernia tends to be wide and the surrounding tissues elastic. Doig and Nixon described a triad of clinical features, namely a reducible buttock swelling, loose stools and an abnormal position of the rectum on digital examination [2] but only the first of these is a consistent finding in all reports. All published cases have been treated by surgical repair. We present a premature infant with bilateral congenital perineal hernias that resolved spontaneously.

2. Case report

A female infant was born by vaginal delivery at 23 weeks' gestation weighing 615 g following spontaneous rupture of membranes. Her mother (gravida 3, para 1) had been stable on a methadone program for 8 years. Two doses of betamethasone were given prior to delivery. The baby was intubated and ventilated for respiratory distress syndrome within minutes of birth but after treatment with surfactant she was rapidly weaned to low pressure volume guaranteed ventilation at low oxygen concentrations. She received a single dose of indomethacin for a patent ductus arteriosus which closed by day 5. A cranial ultrasound scan on day 3 showed an echogenic focus in her left caudate nucleus but subsequent scans were normal. The infant received parenteral nutrition via a peripheral intravenous long line until full enteral feeding with breast milk was established. She was extubated on day 4 and weaned from continuous positive airway pressure support to high flow oxygen by 10 weeks; she was off all respiratory support by 16 weeks of age. Stage I retinopathy of prematurity resolved spontaneously. There were no other complications from her extreme prematurity.

At three weeks of age, she was noted to have bilateral hernias in both buttocks, either side of the anal canal, larger on the right (Figs. 1 and 2). These were confirmed as perineal hernias by ultrasound scan (Fig. 3). Both hernias were easily reducible and were managed conservatively. The left perineal hernia was no longer clinically detectable at the time of discharge home at 17 weeks of age and the right sided hernia was smaller and remained easily reducible. Three weeks later neither hernia was palpable. An MRI scan of the pelvis at 27 weeks of age (equivalent to 10 weeks post term when corrected for prematurity) demonstrated only a subtle outpouching of the distal rectum on the right side that did not extend below the level of the levator ani. At her last review at 15 months of age the baby was thriving and developing normally with no clinical evidence of a perineal hernia.

3. Discussion

Only six cases of congenital perineal hernias presenting in

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Fig. 1. Clinical appearance of bilateral perineal hernias.

Fig. 2. Plain abdominal radiograph showing gas within both perineal hernias. Note that these gas shadows are too medial to represent inguinal hernias.

Fig. 3. Ultrasound scan at 3 weeks of age. Right parasagittal image from a posterior approach (patient prone) showing herniated bowel extending into the ischioanal fossa well below the pelvic floor (short arrows). C = coccyx, Fem = medial aspect of right femoral head.

Table 1
Reported cases of congenital perineal hernia.

No.	Reference	Origin	Sex	Gestation (wks)	Age at diagnosis	Assessment	Clinical features	Associated problems	Treatment	Outcome
1	Moschcowitz (1918)[1]	USA	F	Not stated	2.5y	Clinical	Right buttock swelling - posterior perineal hernia with sac	Extraperitoneal "lipoma"	Surgical repair at 2.5y through buttock incision; defect closed with interrupted sutures reinforced with gluteus maximus	Recovered from surgery. No follow up stated
2	Doig and Nixon (1972)[2]	UK	F	Term	1 m	Barium enema	Left buttock swelling - posterior perineal hernia containing rectum	Intermittent diarrhea	Surgical repair at 11 m; oblique incision over left buttock	No recurrence at follow up but interval not stated
3			M	Term	1 d	Clinical	Right buttock swelling - posterior perineal hernia containing rectosigmoid colon and small bowel	Diarrhea; previous meconium peritonitis	Surgical repair at 3wks; oblique incision over right buttock	Well 2 m later
4	Hubbard and Egelhoff (1989)[3]	USA	F	Not stated	1 d	Plain radiology, barium enema	Right buttock swelling - posterior perineal hernia containing rectosigmoid colon		Surgical repair (no details)	
5	Mohta and Bhargava (2004)[4]	India	F	Term	12 d	Plain radiology, ultrasound, contrast enema, CT	Perineal mass. Right posterior perineal hernia containing rectosigmoid colon	Mild constipation, anus displaced and stenosed by mass (mesenchymal hamartoma)	Surgical repair in neonatal period with excision of perineal mass. Levator ani defect sutured	Well 2y later
6	Kravarusic et al. (2012)[5]	Israel	F	36	4 m (but noticed at birth)	Barium enema, MRI	Left buttock swelling - posterior perineal hernia containing rectum; extraperitoneal with no hernial sac	Mild constipation	Surgical repair at 14 m via posterior sagittal approach; rectal wall repositioned above levator ani and defect sutured, reinforced with gluteus maximus	Uneventful recovery; well 1y later
7	Present case	New Zealand	F	23	2wks	Abdominal radiograph, Ultrasound, MRI	Bilateral buttock swellings - posterior perineal hernias	Extreme prematurity	Conservative	Hernias resolved by 10wks post-term