Staged management of pseudoexstrophy with omphalocele and wide pubic diastasis

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Abstract
Pseudoexstrophy is a rare, mild exstrophy variant characterized by the major musculoskeletal defects of the exstrophy complex without associated defects of the urinary system. The authors present a case of pseudoexstrophy with a small omphalocele and wide pubic diastasis (greater than 4 cm). Only three cases of pseudoexstrophy with omphalocele have previously been described worldwide, and this is the first reported case with wide pubic diastasis. Successful two-stage surgery consisting of omphalocele repair in the neonatal period and abdominoplasty with iliac osteotomy after the age of 6 months is described.

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Classic exstrophy characterized by an exposed bladder plate, open urethra, wide pubic diastases, low umbilicus, and wide rectus has an incidence of approximately 1 in 40,000 births. Exstrophy variants are less common, and usually occur in females [1]. Pseudoexstrophy is a rare, mild exstrophy variant characterized by the major musculoskeletal defects of the exstrophy complex without associated defects of the urinary system [2,3]. Only 17 cases of pseudoexstrophy have previously been reported in the world literature, and pseudoexstrophy with omphalocele and wide pubic diastasis (greater than 4 cm) has not previously been described. We herein report a case of pseudoexstrophy with a small omphalocele and wide pubic diastasis. The management of pseudoexstrophy with wide pubic diastasis is discussed.

1. Case report

A 2774 g infant girl was born by vaginal delivery following an uncomplicated term gestation. Physical examination revealed a low-set umbilicus with a small omphalocele and bifid clitoris (Fig. 1A). The anus was anteposed. Cardiac and cranial ultrasound scans were normal. Chromosomal analysis showed a normal 46, XX karyotype. Abdominal X-ray showed separation of the pubic symphysis with outward rotation of the innominate bones (Fig. 1B). As the pubic diastasis was greater than 4 cm, pelvic osteotomy would be needed to repair the abdominal wall and pubic diastasis. However, the pelvic ring was not strong enough to repair and there were a risk of excessive intra-abdominal and pelvic pressure. Hence, the omphalocele was repaired on day 2 of life and abdominoplasty with pelvic osteotomy was planned after the age of 6 months. During repair of the abdominal wall defect, the presence of a normal bladder in the hernia was confirmed. The postoperative recovery was uneventful, and the patient was discharged on day 14 of life. Computed tomography at 7 months of age showed pubic diastasis of 6 cm. At 8 months of age, abdominoplasty was undergone with iliac osteotomy. Bilateral posterior iliac osteotomy was performed in the prone position. The pubic rami were exposed by subperioisteal dissection in the supine position, and were secured in position by passing a Leeds-Keio® artificial ligament (Xiros, Leeds, UK) through the obturator foramina. On each side, the inner half of the rectus muscles was detached from the pubic bone and then fixed to the contralateral part of the pubic bone. The rectus muscles were approximated in layers without tension and the umbilicus was moved to a more normal position. The postoperative recovery was uneventful. The patient was mobilized with a pelvic belt for 12 weeks. At the 7-month follow-up, she was asymptomatic and there was no recurrence of pubic diastasis (Fig. 2A, B).
2. Discussion

Pseudoexstrophy is the mildest exstrophy variant and is characterized by a low-set umbilicus, lower rectus diastasis, and pubic symphysis abnormality. The term pseudoexstrophy was first used in 1954 by Hejtmancik et al. [2]. Although their patient presented with severe bladder prolapse between divergent abdominal muscles, the bladder was intact and there was no evidence of exstrophy [2]. The patient underwent surgical repair with mesh reinforcement, and the postoperative recovery was uneventful. Seventeen cases of pseudoexstrophy have previously been reported, of which 14 cases required surgical intervention [2–6]. Only three cases of pseudoexstrophy associated with omphalocele have been reported worldwide and pseudoexstrophy with omphalocele and wide pubic diastasis (greater than 4 cm) has not previously been reported [4–6]. There is no standard surgical strategy for pseudoexstrophy with wide pubic diastasis.

In patients with bladder exstrophy who have wide pubic diastasis (greater than 4 cm), are older than 72 h, or are undergoing bladder closure, concurrent osteotomy increases the reliability of pelvic ring closure [7]. Pelvic osteotomy and postoperative immobilization have been shown to decrease the failure rate after repair of bladder exstrophy [7]. Children who were older at the time of the osteotomy were reported to maintain better correction over time [8]. It is recommended that the procedure should be delayed until at least 6 months of age, and until the pelvic ring is strong enough to place an intrasymphseal device [9]. Early correction of the pelvic anatomy may enable normal bone growth, as there is evidence that bony development depends on exposure to mechanical forces [10]. However, one-stage primary closure in the neonatal period may cause excessive intra-abdominal and pelvic pressure, which may result in abdominal compartment syndrome and respiratory problems. Moreover, rapid manipulation of the pelvic anatomy may lead to femoral nerve palsy. If the diastasis is 4 cm or less, one-stage primary closure in early infancy or childhood may be suitable [9]. However, if the diastasis is greater than 4 cm, staged abdominoplasty and osteotomy may be a safer option for patients with pseudoexstrophy.

Our patient had pseudoexstrophy with a small omphalocele and wide pubic diastasis. The pubic diastasis was greater than 4 cm, and pelvic osteotomy was needed to repair the abdominal wall and pubic diastasis. We performed two-stage repair, consisting of omphalocele repair in the neonatal period and abdominoplasty with iliac osteotomy after the age of 6 months. The postoperative recovery was uneventful for both procedures, and there was no recurrence of the pubic diastasis.

3. Conclusion

This is the first report of pseudoexstrophy with a small omphalocele and wide pubic diastasis. Staged management may be a safe and effective option for the patients with this association.

Disclosures

The authors declare that they have no relevant financial interests. All authors reviewed and approved the manuscript.
References


