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Challenges in the management of a rare case of caudal duplication syndrome in a poor resource setting

E. Bandré^{a,*}, A. Wandaogo^a, S.M. Kabre^b, I. Ouedraogo^a, M. Napon^c^a Department of Paediatric Surgery, Charles de Gaulle Paediatric Teaching Hospital, Ouagadougou, Burkina Faso^b Department of Radiology, Charles de Gaulle Paediatric Teaching Hospital, Ouagadougou, Burkina Faso^c CMA Schiffr, Ouagadougou, Burkina Faso

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ABSTRACT

Caudal duplication syndrome is a series of malformations affecting the development of the caudal area of the embryo involving a combination of malformations of the digestive tract, the genitourinary tract, the spinal column, the limbs or the neural tube. The authors report a case characterized by a supernumerary lower limb comprised of a thigh, leg, two feet joined by their medial edge and two scrota each containing one testicle and two phalluses, one of which lacks a urethral. The second phallus had an apical urethral meatus allowing for normal urination, a hemi-thoracic vertebrae, a megaureter and a single kidney and supernumerary vertebrae. A surgical excision of the supernumerary limb and the abnormal phallus was performed, followed by a fusion of the two scrota. The surgical outcomes were uneventful.

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Reported for the first times in ‘Ephemerides’ of Leopoldine Academy at Frankfurt from 1712 to 1717 [1]; an adequate description of caudal duplication syndrome was provided by Dominguez et al. in 1993. It is a rare polymalformative entity affecting the limbs, the genitourinary tract, the digestive tract, the neural tube and the spinal column. It consists of a duplication of the genitourinary system, the gastrointestinal system, the spinal column, the spinal cord and the lower limbs [2].

We report a case diagnosed at the age of 3 months in order to illustrate the procedures for its treatment in developing countries.

1. Case report

S.A.I. was a male newborn. He was received at the Charles De Gaulle Teaching Paediatric Hospital Centre at Ouagadougou at three months for supernumerary limb. In the family history, he was the fifth child of the uterine siblings of 5 children. The mother was 42

years old and the father 52 years old. There was no consanguinity between parents. No prenatal consultation was made during the pregnancy. The mother naturally delivered at term at home. The clinical examination showed an infant with an overall good condition, weighing 6 kg, a lower supernumerary limb comprising a thigh, a leg, two feet joined together by their medial edge with 10 toes, two scrota, each containing one testicle and two phalluses. The right phallus measured 4 cm with a scrotum measuring 2 cm in diameter, containing a normal testicle without a urethral canal and without a urethral meatus. The second phallus, the left measured 3 cm with a scrotum measuring 2 cm in diameter containing a normal testicle. This phallus had a urethra terminating in an apical urethral meatus allowing for normal urination (Fig. 1). The sensitivity and motor function of the supernumerary limb was retained. The diagnosis of caudal duplication syndrome was proposed. The CT scan of the chest, abdomen and pelvis and the CT scan of the limbs revealed:

- in the supernumerary limb: a femur articulating by a neo-articulation between the two ischia, the two tibiae and the tarsals and metatarsals of the joined feet (Fig. 2).
- thoracic scoliosis with 13 thoracic vertebrae including one hemi-vertebrae in the 10th thoracic vertebrae (Fig. 3).
- 12 ribs in the right and 11 ribs in the left (Fig. 3).

* Corresponding author. Department of Paediatric Surgery, Charles de Gaulle Paediatric Teaching Hospital 01, P.O. Box 1198, Ouagadougou 01, Burkina Faso. Tel.: +226 70239578.

E-mail address: brice52001@yahoo.fr (E. Bandré).



Fig. 1. Supranumerary lower limb, diphallus, two scrota.

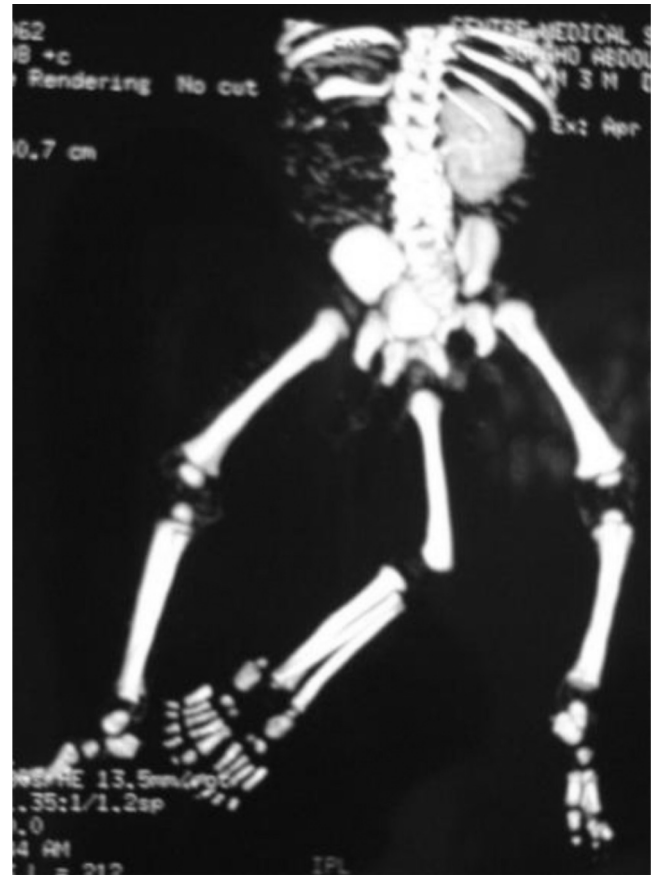


Fig. 2. CT scan frontal view showing a femur articulating by a neo-articulation between the two ischia, the two tibiae and the tarsals and metatarsals of the joined feet.

The abdominal ultrasound and the CT urography revealed a single functional large right kidney and a megaureter draining the urine to a normal bladder.

There is no colonic or rectal duplication in the barium enema.

Surgery performed consisted in an excision of the supranumerary limb, the non functional supranumerary phallus and a plasty of the scrotum by fusion of the two scrota to form one scrotum (Fig. 4).

The surgical outcomes were simple. The patient was seen every three months to monitor his malformative uropathy and his vertebral malformation while awaiting future surgical correction of his scoliosis.

At three years of age (30th September 2015), the surgical outcome was always uneventful. There is no dysurie. Cosmetic result of the scrotum is acceptable (Fig. 5). Clinically there is no curvature of spine in the lateral plane (Fig. 6) and no limp or complain in the walking (Fig. 7).

2. Discussion

Caudal duplication syndrome is a rare anomaly, reflected by the small number of cases reported in the literature [1]. Certain sociological and cultural pressures [3] still present in Burkina Faso result in a tendency to eliminate or hide newborns with severe birth defects, which explains the long delay in bringing this infant to the hospital.

The exact etiology and pathogenesis of caudal duplication syndrome are unknown. Several theories have been proposed:

- Bajpai et al. [4] have been suggested that this constellation of anomalies endorse the theory of caudal twinning as the mechanism behind caudal duplication.

- Pang et al. [5] have postulated an abnormal adherence between ectoderm and endoderm.
- For Dominguez et al. [2] the causative factor of caudal duplication syndrome is the damage to caudal cell mass to the 23–25 day embryo.
- Al Alayet et al. [6] conclude that the occurrence of these constellations of anomalies typical of caudal duplication is the result of disruption of regulation of WNT pathway and possibly involving Axin1 gene at the early part of embryogenesis.

The lack of prenatal diagnosis did not allow a diagnosis, monitor this pregnancy monthly which may get complicated, inform parents of the anomaly found and organize the delivery in a health facility equipped with a surgery unit. This weakness has led to late diagnosis, the lack of psychological preparation of parents, and delivery in inappropriate conditions.

Despite the lack of obstetric complications in the mother who delivered naturally, the resort to caesarean should be systematic to prevent a possible obstructed labor. After delivery, the medical pediatric and surgery care should be organized as soon as possible.

This case is different of those generally reported by others authors in which the malformations concerned also duplication of caudal vertebral column with meningocele, imperforate anus, colonic rectal duplication, double bladder [2,4,8] or only the limbs [7].

Treatment of these anomalies consists of staged correction. Corrective surgery by fusion of the two scrota and resection of the non functional supranumerary phallus and the



Fig. 3. Thoracic-abdominal antero-posterior X-ray: showing thoracic scoliosis with 13 thoracic vertebrae including one hemi-vertebrae in the 10th thoracic vertebrae; 12 ribs in the right and 11 ribs in the left.



Fig. 5. Cosmetic result of the scrotum.



Fig. 4. Post operative aspects after excision of supernumerary lower limb, non functional phallus, fusion of the two scrota.



Fig. 6. Clinical aspect on posterior view of the patient.



Fig. 7. Clinical aspect on frontal view of the patient.

supernumerary member was done successfully. Corrective of the scoliosis will be done after. The management of this polymalformative syndrome must be performed step by step by a

multidisciplinary team of radiologist, orthopedic pediatricians, neurosurgeons and urologists not yet available in our country, which suffers from limited resources. The emphasis in the treatment of these malformations must be placed first on the malformations which are potentially life threatening or the most crippling for the child.

3. Conclusion

The treatment of caudal duplication syndrome is particularly difficult in a developing country with limited resources. To improve the prognosis of these anomalies, which require systematic investigation, multidisciplinary treatment, a sufficient number of qualified personnel must be trained, surgical units must be created and equipped and the major congenital defects must be included in the national programs for integrated treatment of child and maternal diseases.

Conflict of interest

No conflict of interest.

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