

## Cloacal Exstrophy: Documenting a Particular Case in Yaoundé, Cameroon

**Kamsu Zicfried\*, Aurore Albane Essomba, Laura Kuate Makowa, Raïssa Monayong Mendomo, Sonia Zambou Zebaze, Audrey Thérèse Mbang, Christiale Batibonak and Oummy Djamila Ngapout**

*Faculty of Medicine and Biomedical Sciences, The University of Yaoundé 1, Yaoundé, Cameroon*

\*Corresponding Author

**Kamsu Zicfried**

**Abstract:** Cloacal exstrophy in its most complex form with OEIS syndrome is characterized by the existence of an Omphalocele, Exstrophied bladder, Imperforate anus and Spina bifida. We report a particular case of Cloacal exstrophy with OEIS complex and further birth defects. The diagnostic and therapeutic approaches, as well as difficulties encountered in a limited resource setting are highlighted. This was a 1-day old neonate referred for the management of multiple congenital malformations, including antenatal diagnosis of malformative uropathy. On admission, the clinical findings included: a type 1 omphalocele, an anorectal malformation with a recto-urinary fistula and a covered lumbosacral dysraphism. Paraclinical examinations with cardiac and transfontanelle ultrasound revealed associated cardiac anomaly and findings in favor of lombo-sacral dysraphism. Supportive care was given and surgical reconstruction of birth defects on day 20 was done. In conclusion, cloacal exstrophy is a rare morbid congenital polymalformative syndrome in neonates, especially when presenting with OEIS complex or syndrome. They require prompt diagnosis and immediate postnatal multidisciplinary management, with long-term follow-up for a favorable outcome.

**Keywords:** Cloacal exstrophy, OEIS complex, congenital malformation, polymalformative syndrome

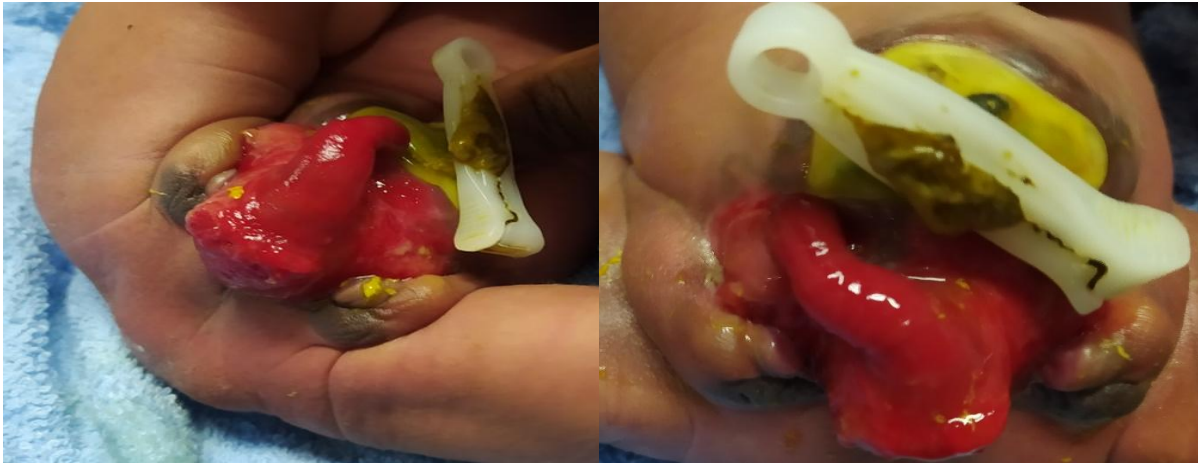
### INTRODUCTION

Cloacal exstrophy in its most complex form with OEIS syndrome is characterized by the existence of an Omphalocele, Exstrophied bladder, Imperforate anus and Spina bifida. These are grouped under the term OEIS complex or syndrome [1, 2]. It is often associated with other malformations, involving the gastrointestinal and skeletal spheres [1]. It was first described in 1709 by LITTRE, and for a long time was considered a devastating pathology due to infectious, metabolic or nutritional complications associated with short bowel syndrome and intestinal obstructions [3, 4]. In effect, the prognosis, although dependent of associated anomalies, remains uncertain in context with low technical platform.

### Case presentation

This is a newborn baby on its first day of life referred for the management of a congenital malformation noted at birth. The mother was primiparous, aged 22 years, there wasn't a notion of consanguinity. Four antenatal consultations had been carried out in a remote rural area with an undocumented infectious work-up. We did not find any exposition of teratogenic substances. There was no history of chronic or pregnancy related pathology in the mother. She had received iron and folic acid throughout the pregnancy in adequate doses. A 33 weeks' gestation obstetrical ultrasound had showed pyelocal dilatation of the right kidney in favor of fetal uropathy. Delivery was per vaginal at 35 weeks' gestation + 3 days, with amniotic fluid described as meconium stained and fetid. The birth weight was 2000 g with an Apgar score of 8 and 10 at the 1st and 5th minute respectively.

Physical examination on admission revealed a newborn with normal vital and anthropometric parameters for gestational age. Signs of prematurity with estimated gestational age of 36 weeks according to the Ballard score. We had a type 1 omphalocele with umbilical hernia (fig1), cloacal exstrophy with two exstrophied hemi bladders, separated with everted cecum (fig1). There was a sexual differentiation anomaly, as well as anorectal malformation with recto-urinary fistula (fig2) and a covered lumbosacral dysraphism (fig3).



**Figure 1 and 2:** omphalocele type 1, cloacal exstrophy, anal imperforation, external genitalia ambiguity



**Figure 3:** spina bifida occulta meningocele

Cardiac ultrasound revealed an ostium secundum type inter-atrial communication of approximately 2.5 cm, the interventricular septum being intact (fig4). Abdominal and pelvic ultrasound concluded there was bladder exstrophy, with integrity of the intra-abdominal organs. The transfontanelar ultrasound showed a posterior superficial formation of pure anechoic content measuring 38 x 17 x 33 mm, that is 11 cc in volume, with an opening into the medullary canal, suggesting a meningocele. We requested karyotype, which could not be done due to lack of financial means.

The newborn was put on triple antibiotic therapy (cefotaxim, ampicillin and gentamycin) in view of the meconium-stained and fetid coloration of the amniotic fluid for 10 days according to the department's protocol. Enteral feeding with breast milk was started on the second day of hospitalization due to the absence of secretions coming back through the nasogastric tube. Twice daily wet dressings of the abdomen were performed until the surgery.

The child underwent surgery on the 20th day of life. This consisted of a two-step surgical intervention, of which the first comprised an exploratory laparotomy with a peri omphalocele incision and the reconstruction of digestive and uro-genital defects. The second step consisted of lumbosacral dysraphism meningocele repair.



**Figure 4:** Ostium secundum type atrial septal defect of about 2.5 cm

## DISCUSSION

Cloacal exstrophy is an extremely rare polymalformative syndrome with prevalence estimation as low as to 1/400000 live births. It is believed to be predominant in male neonates when the gonads and external genitalia can be well appreciated. This was not the case with our patient due to sexual differentiation anomalies. As the diagnosis of this syndrome is difficult to pose, the very low global incidence reported is thought to be an underestimation by some authors. However, to the best of our knowledge, no such cases had earlier been documented in our context[5-7].

Although a karyotype could not be done in our situation, young maternal age, just as fetid and meconium stained amniotic fluid as infection criteria, were non negligible risk factors for congenital malformations [8-11]. On the other hand, the antenatal ultrasound-based diagnosis of malformative uropathy was a pertinent finding of pregnancy follow-up, which was in favor of Cloacal exstrophy. Early referral to specialized structures for a better management was preconized, but not honored. This was probably because of parental psychological unpreparedness to accept the antenatal diagnosis [12-19]. As a matter of fact, antenatal diagnosis when accepted by parents, offers the advantage of better understanding of the condition, buys time for financial and moral preparation for treatment, makes in utero transfer possible and enables preparation to maternity[5, 19-22]. The clinical diagnosis of cloacal exstrophy in this case was not obvious at birth, as extra malformations were confusing, with a necessity for literature review.

The management of the pathology was multidisciplinary including neonatal resuscitation, supportive care, antibiotherapy and finally surgery. This was planned by taking into consideration the urgencies presented by the patient, given visceral exposure with risk of hydro-electrolytic imbalances, super infection, necrosis and multi visceral failure. Surgery was only practice once the neonate was safe enough, contrary to 72 hours' recommendation in neonatal surgical emergencies[23-28].

The surgical interventions consisted of a patient-adapted protocol with successive reconstruction of the genitourinary and anorectal tracts, followed by omphalocele repair in a first stage. While the second stage intervention consisted of meningoplasty. The interventions were successful and no immediate post-operative complications were noted.

## CONCLUSION

Cloacal Exstrophy is a rare morbid congenital polymalformative syndrome especially when presenting with the OEIS complex. We reported a particular case in our setting associated with further defects which may render diagnosis difficult, with a need for literature review. Regular and effective antenatal follow-up with obstetrical ultrasound had contributed to early antenatal diagnosis. However, psychological unpreparedness of parents was an obstacle for referral and prompt intervention. The management of the neonate at birth was multidisciplinary and interventions were adapted to immediate patient's medical needs. This stabilized and made the patient operable before surgical reconstruction could be practiced, and the outcome was favorable.

## REFERENCES

1. Woo, L. L., Thomas, J. C., & Brock, J. W. (2010). Cloacal exstrophy: a comprehensive review of an uncommon problem. *Journal of pediatric urology*, 6(2), 102-111.
2. Manassero-Morales, G., Franco-Bustamante, K., & Matos-Rojas, I. (2016). OEIS complex, a case report. *J Intensive & Crit Care*, 2, 1.
3. Gbenou, A. S., Assan, B. R., Akodjenou, J., Enianloko, N. T., Yasségoungbé, M. G., & Fiogbe, M. A. (2020). Oeis Syndrome (Cloacal Exstrophy): About Two Cases Treated at the Mother and Child Teaching Hospital in Cotonou (Benin). *Open Journal of Pediatrics*, 10(3), 535-541.
4. Arteaga-Vázquez, J., Luna-Muñoz, L., Morales-Suárez, J. J., & Mutchinick, O. M. (2019). OEIS complex: Prevalence, clinical, and epidemiologic findings in a multicenter Mexican birth defects surveillance program. *Birth Defects Research*, 111(11), 666-671.
5. Gbenou, A. S., Assan, B. R., Akodjenou, J., Enianloko, N. T., Yasségoungbé, M. G., & Fiogbe, M. A. (2020). Oeis Syndrome (Cloacal Exstrophy): About Two Cases Treated at the Mother and Child Teaching Hospital in Cotonou (Benin). *Open Journal of Pediatrics*, 10(3), 535-541.
6. Diamond, D. A., & Jeffs, R. D. (1985). Cloacal exstrophy: a 22-year experience. *The Journal of urology*, 133(5), 779-782.
7. Boyadjiev, S. A., Dodson, J. L., Radford, C. L., Ashrafi, G. H., Beaty, T. H., Mathews, R. I., ... & Gearhart, J. P. (2004). Clinical and molecular characterization of the bladder exstrophy-epispadias complex: analysis of 232 families. *BJU international*, 94(9), 1337-1343.
8. Lubala, T. K., Shongo, M. Y., Mbuyi, S. M., Mutombo, A. M., Ngwej, D. T., & Kabange, F. N. (2013). OEIS Complex (omphalocele-exstrophy of the bladder-anal imperforation-spina bifida) and prenatal alcohol exposure: a case report. *Pan African Medical Journal*, 15(1).

9. Georges Pius K M, Aurore Albane E, Marie-Paul B, Komba D, NgandoVK, Eteme A et al(2022). Neonatal Sepsis: Highlights and Controversies. *J Pediatr Neonatal*, 4(1): 1-5. Doi: 10.33425/2689-1085.1035
10. Chiabi, A. (2019). Relevance and Applicability of the Apgar Score in Current Clinical Practice. *EC Paediatrics*, 8, 1-7.
11. Moyo, G. P. K., & Tetsiguia, J. R. M. (2020). Discussing the “First Cry” as an Initial Assessment for Neonates. *American Journal of Pediatrics*, 6(2), 129-132.
12. Sowmya M, Shwetha S.(2014). Oeis Complex: a rare Case Report. *Int J Med Res Rev*, 2(6):618-620.
13. Chou, C. Y., Tseng, Y. C., & Lai, T. H. (2015). Prenatal diagnosis of cloacal exstrophy: a case report and differential diagnosis with a simple omphalocele. *Journal of Medical Ultrasound*, 23(1), 52-55.
14. Moyo, G. P. K., & Djoda, N. (2020). The Emotional Impact of Mode of Delivery in Cameroonian Mothers: Comparing Vaginal Delivery and Caesarean Section. *American Journal of Psychiatry and Neuroscience*, 8, 22-25.
15. Moyo, G. P. K., Mendomo, R. M., Zebaze, S., Makowa, L. K., Batibonack, C., & Mbang, A. T. (2020). Neonatal Determinants of Mothers’ Affective Involvement in Newly Delivered Cameroonian Women. *American Journal of Pediatrics*, 6(2), 125-128.
16. Moyo, G. P. K. (2021). Perinatal and Childbirth as a Factor of Decompensation of Mental Illness: The Case of Depressive States in Newly Delivered Cameroonian Women. *Arch Med*, 7(2).
17. Moyo, G. P. K., & Djoda, N. (2020). Relationship Between the Baby Blues and Postpartum Depression: A Study Among Cameroonian Women. *American Journal of Psychiatry and Neuroscience*, 8(1), 26-29.
18. Moyo, G. P. K. (2020). Epidemio-clinical Profile of the Baby Blues in Cameroonian Women. *Journal of Family Medicine and Health Care*, 6(1), 20-23.
19. Foumane, P., Olen, J. P. K., Fouedjio, J. H., Moyo, G. P. K., Nsahlai, C., & Mboudou, E. (2016). Risk factors of maternity blues after caesarean section in Yaoundé, Cameroon: a case-control analysis. *International Journal of Reproduction, Contraception, Obstetrics and Gynecology*, 5(12), 4424-4428.
20. Hermann, N. D. (2020). Neonatal Determinants of Inadequate Breastfeeding: A Survey among a Group of Neonate Infants in Yaoundé, Cameroon. *Open Access Library Journal*, 7(07), 1.
21. Dany, H. N., Moyo, G. P. K., Ejake, L., Nguéack, F., Mah, E., Maguip, L., ... & Chiabi, A. (2020). Determinants of Breastfeeding Initiation Among Newly Delivered Women in Yaounde, Cameroon: a Cross-Sectional Survey. *HEALTH SCIENCES AND DISEASE*, 21(9).
22. Moyo, G. P. K., & Hermann, N. D. (2020). Clinical Characteristics of a Group of Cameroonian Neonates with Delayed Breastfeeding Initiation. *American Journal of Pediatrics*, 6(3), 292-295.
23. Moyo, G. P. K., & Hermann, N. D. (2020). The Psycho-Sociocultural Considerations of Breastfeeding in a Group of Cameroonian Women with Inadequate Practices. *Journal of Psychiatry and Psychiatric Disorders*, 4(4), 130-138.
24. Moyo, G. P. K., Ngwanou, D. H., Sap, S. N. U., Nguéack, F., & Mah, E. M. (2020). The Pattern of Breastfeeding among a Group of Neonates in Yaoundé, Cameroon. *International Journal of Progressive Sciences and Technologies*, 22(1), 61-66.
25. Varygin, V., Bernotas, Š., Gurskas, P., Karmanovas, V., Strupas, S., Zimanaitė, O., & Verkauskas, G. (2011). Cloacal exstrophy: a case report and literature review. *Medicina*, 47(12), 100.
26. Ngwanou, D. H., Ngantchet, E., & Moyo, G. P. K. (2020). Prune-Belly syndrome, a rare case presentation in neonatology: about one case in Yaounde, Cameroon. *Pan African Medical Journal*, 36(1).
27. Moyo, G. P. K., Nguedjam, M., & Miaffo, L. (2020). Necrotizing Enterocolitis Complicating Sepsis in a Late Preterm Cameroonian Infant. *American Journal of Pediatrics*, 6(2), 83-86.
28. Moyo, G. P. K., Sobguemezing, D., & Adjifack, H. T. (2020). Neonatal Emergencies in Full-Term Infants: A Seasonal Description in a Pediatric Referral Hospital of Yaoundé, Cameroon. *American Journal of Pediatrics*, 6(2), 87-90.