

## Case report

### **OEIS Complex (omphalocele-exstrophy of the bladder-anal imperforation-spina bifida) and prenatal alcohol exposure: a case report**

**Toni Kasole Lubala<sup>1</sup>, Mick Yapongombo Shongo<sup>1,2</sup>, Sébastien Musanzayi Mbuyi<sup>1,2</sup>, Augustin Mulangu Mutombo<sup>1</sup>, Dieudonné Tshikwey Ngwej<sup>1</sup>, Félix Numbi Kabange<sup>1</sup>**

<sup>1</sup>Faculté de Médecine, Université de Lubumbashi, 1825 Lubumbashi, République Démocratique du Congo, <sup>2</sup>Centre Interdisciplinaire de Génétique au Congo, CIGEC, Lubumbashi, République Démocratique du Congo

<sup>8</sup>Corresponding author: Toni Kasole Lubala, Faculté de Médecine, Université de Lubumbashi, 1825 Lubumbashi, République Démocratique du Congo

Key words: Omphalocele, exstrophy of the bladder, anal imperforation, spina bifida, alcohol

Received: 07/12/2012 - Accepted: 06/07/2013 - Published: 20/08/2013

#### **Abstract**

We describe a Congolese case of OEIS complex: Omphalocele-Exstrophy of the Bladder-anal imperforation-spina bifida, rare congenital malformation complex. Its genetic and environmental factors are not well known. In our case, the fetus has been exposed to alcohol throughout the all pregnancy and the mother contracted a gonococcal infection during the two first trimesters of her pregnancy.

**Pan African Medical Journal. 2013; 15:142. doi:10.11604/pamj.2013.15.142.2256**

This article is available online at: <http://www.panafrican-med-journal.com/content/article/15/142/full>

© Toni Kasole Lubala et al. The Pan African Medical Journal - ISSN 1937-8688. This is an Open Access article distributed under the terms of the Creative Commons Attribution License (<http://creativecommons.org/licenses/by/2.0>), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

## Introduction

---

The OEIS Complex describes a rare grouping of more commonly occurring component malformations first describe by Carey et al. in 1978 [1,2]. The etiology is unknown, but likely heterogeneous. Its incidence is estimated at 1 for 200000 live births in Europe [3] and 1 for 400000 in Australia [4]. Some authors suggested that it is the most severe form of the "Exstrophy of the bladder-Epispadias complex" which has a large clinical spectrum from isolated epispadias to the OEIS complex [3,5]. If, on one hand, many authors present evidences of the existence of genetic factors in the OEIS complex, the environmental factors that can contribute to the development of this congenital complex of malformations on the other hand, are still unknown. We describe, in this article, a case of OEIS complex in a Congolese new born baby who was exposed to alcohol in utero.

## Patient and observation

---

We report on a male, full term new born of normal delivery born at a maternity clinic of the Katanga province, southern Democratic Republic of Congo. The father is a 40 year old business man and the mother a 31 year old house wife. Her past obstetric history revealed a previous spontaneous miscarriage at 12 week during the second pregnancy (**Figure 1**). The new born reported about in this article is the seventh child of the couple and nothing specific was noted on the other siblings. Their marriage is not consanguineous. The mother only had only two antenatal consultations where a gonococcal infection was diagnosed. None obstetrical ultrasound was performed during the pregnancy neither did the mother receive the folic acid supplementation normally require during any pregnancy. The mother mentioned a regular intake of at least one glass of beer (4% of alcohol) daily before and during the pregnancy but no notion of tobacco consumption. In the labor ward a hydramnios was noted. The clinical examination revealed a hypotrophy with a low birth weight of 2380g (-2SD -3SD), a length of 44cm (-3SD), a thorax circumference of 31 cm and a microcephaly with a head circumference of 32 cm (-2SD -3SD). A medial sub-umbilical closure defect of the anterior abdominal wall was observed with omphalocele, vesical exstrophy (**Figure 2**) and imperforate anus. Was also noted an emission of the meconium through the urethral meatus which is the sign of a recto-vesical fistula. There was no epispadias. The examination of the spinal

column revealed a spina bifida occulta (**Figure 3**). The cardiac and abdominal ultra sounds (performed to search for possible heart or kidney malformation) were normal. The HIV and VDRL serology were negative.

## Discussion

---

The OEIS complex is a rare congenital malformation whose etiological factors are not well known up to now. The sporadic nature of most of the reported cases, however may suggest multiple etiologic factors as well as a genetic and environmental interaction [6]. Authors of some descriptive studies suggested that the age of the parents, the higher parity and the race might be predisposing factors [3]. In another large study on 186 patients chosen from congenital malformation registers of 24 different countries though, the incidence did not vary with the maternal age [7]. A vesical exstrophy was found in association with congenital rubella. This malformation was also found in fetus that had been exposed to teratogenic substances like Diazepam and alcohol [3, 8].

The mother contracted a gonococcal infection at the beginning of the pregnancy. It is the first case of OEIS complex appearing in this context. The possibility of a gonococcal infection being an etiological factor of OEIS complex has to be study further. We note moreover, a fetal alcohol exposure throughout the all pregnancy. Even though the newborn did not present the craniofacial abnormalities that characterize the fetal alcohol syndrome, the harmonious hypotrophy and the microcephaly observed can be due to alcohol exposure of the fetus [9]. We have to acknowledge though, that the link between alcohol exposure in utero and the occurrence of the OEIS complex cannot be established at this point.

Regarding the genetic aspects, the newborn was the first case of OEIS Complex observed in the family. However, it is interesting to note that the mother is the only one alive in a family of five children; early neonatal death, of cause unknown by the mother, occurred for the other siblings.

A case report of OEIS complex has been associated with a 3q 12.2-13.2 deletion and more recently, another case was associated with a monosomy 1p36 deletion [10, 11]. However, further research need to be done in order to determine whether or not, these structural

chromosome abnormalities could be etiological factors of the OEIS complex.

In some studies reporting cases of recurrence in families, at least two different members of the same family were affected at different degree [7]. In two families for instance, it was two members of different sex but in a Moroccan family, three different cases of OEIS complex were observed on male, suggesting an X-linked transmission [7]. Literature also mentioned the possibility of an association between a new dominant mutation which might occur in one of the two parental gonads and the expression of an OEIS complex [12].

The surgical treatment is difficult due to the complexity of this grouping of malformations; it often offers poor results and does not spare the patient from the psychological consequences of any major urogenital surgery. In developing countries like ours, this complex is still of very poor prognosis.

## Conclusion

---

We describe a new case of Omphalocele-vesical exstrophy-Imperforate anus-spina bifida complex (OEIS Complex) on a male newborn who was exposed to alcohol in utero and whose mother contracted a gonococcal infection during the two first trimesters of the pregnancy.

## Competing interests

---

The authors declare no competing interests.

## Authors contributions

---

Toni Kasole Lubala: Redaction of the manuscript, diagnosis, photography. Nina Lubala: Diagnosis, Review of the literature and manuscript English translation. Arthur Ndundula Munkana: Management of pregnancy and antenatal diagnosis. Augustin Mulangu Mutombo: Pediatric care. Mick YaPongombo Shongo: Pediatric care. Sebastien Musanzayi Mbuyi: Surgical Management. Dieudonné Tshikwej Ngwej: Correction of the manuscript. Félix

Numbi Kabange: Correction of the manuscript. All the authors have read and approved the final version of the manuscript

## Figures

---

**Figure 1:** The past obstetric history of the patient revealed a previous spontaneous miscarriage at 12 week during the second pregnancy

**Figure 2:** A medial sub-umbilical closure defect of the anterior abdominal wall was observed with omphalocele, vesical exstrophy and imperforate anus

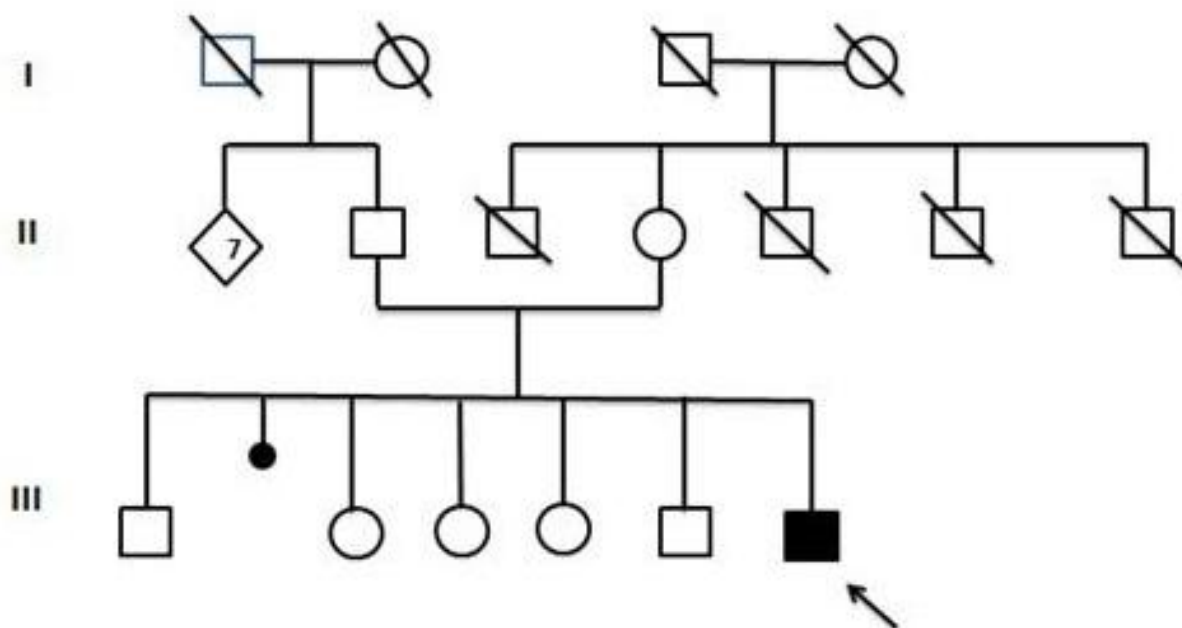
**Figure 3:** The examination of the spinal column revealed a spina bifida occulta

## References

---

1. Varygin V, Bernotas S, Gurskas P, Karmanovas V, et al. Cloacal exstrophy: a case report and literature review. *Medicina*. 1978; 47(12): 682-5. **PubMed** | **Google Scholar**
2. Carey JC, Greenbaum B, Hall BD. The OEIS complex (omphalocele, exstrophy, imperforate anus, spinal defects). *Birth Defects Orig Artic Ser*. 1978; 14: 253-63. **PubMed** | **Google Scholar**
3. Anne-Karoline Ebert, Heiko Reutter, Michael Ludwig, Wolfgang H Rösch. The Exstrophy-epispadias complex. *Orphanet J Rare Dis*. 2009 Oct 30;4:23. **PubMed** | **Google Scholar**
4. Smith NM, Chambers HM, Furness ME, et Haan EA. The OEIS complex (omphalocele-exstrophy-imperforate anus-spinal defects): recurrence in sibs. *J Med Genet*. 1992; 29(10): 730-2. **PubMed** | **Google Scholar**
5. Martínez-Frías ML, Bermejo E, Rodríguez-Pinilla E, Frías JL. Exstrophy of the cloaca and exstrophy of the bladder: two different expressions of a primary developmental field defect. *Am J Med Genet*. 2001; 99(4): 261-69. **PubMed** | **Google Scholar**

6. Keppler-Noreuil KM. OEIS complex (omphalocele-exstrophy-imperforate anus-spinal defects): a review of 14 cases. *Am J Med Genet.* 2001; 99(4): 271-9. **PubMed| Google Scholar**
7. Feldkamp M, Botto LD, Amar E, Bakker MK, et al. Cloacal exstrophy: an epidemiologic study from the international Clearinghouse for Birth Defects Surveillance and Research. *Am J Med Genet C Semin Med Genet.* 2011; 157(4): 333-43. **PubMed| Google Scholar**
8. Lizcano-Gil LA, García-Cruz D, Sánchez-Corona J. Omphalocele-exstrophy-imperforate-anus-spina bifida (OEIS) complex in a male prenatally exposed to diazepam. *Arch Med Res.* 1995; 26(1): 95-6. **PubMed| Google Scholar**
9. Bernard Dan et Yves Gillerot. Le syndrome d'alcoolisme foetal. Education du patient et enjeux de santé. 2005; 23(3): 70-2. **PubMed| Google Scholar**
10. Ben-neriah Z, Withers S, Thomas M, Toi A, et al. OEIS complex: prenatal ultrasound and autopsy findings. *Ultrasound Obstet Gynecol.* 2007; 29: 170-177. **PubMed| Google Scholar**
11. Kosaki R, Fukuhara Y, Kosuga M, Okuyama T, et al. OEIS complex with del(3)(q12.2q13.2). *Am J Med Genet A.* 2005 Jun 1;135(2):224-6. **PubMed| Google Scholar**
12. El-Hattab AW, Skorupski JC, Hsieh MH, Breman AM, et al. OEIS complex associated with chromosome 1p36 deletion: a case report and review. *Am J Med Genet A.* 2010 Feb;152A(2):504-11. **PubMed| Google Scholar**



**Figure 1:** The past obstetric history of the patient revealed a previous spontaneous miscarriage at 12 week during the second pregnancy



**Figure 2:** A medial sub-umbilical closure defect of the anterior abdominal wall was observed with omphalocele, vesical exstrophy and imperforate anus



**Figure 23:** The examination of the spinal column revealed a spina bifida occulta

