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Case report

# A rare presentation of OEIS variant with a recto-bladder neck fistula: A case report and literature review

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ABSTRACT

*Introduction:* Omphalocele, bladder extrophy, imperforate anus and spinal defect (known as OEIS) is a very rare congenital anomaly with an unknown etiology. In this report we describe a case of an OEIS variant associated with a wide pubic diastasis, bladder extrophy with a recto-bladder neck fistula and a high ano-rectal malformation. This work has been reported in line with the SCARE 2020 criteria.

*Presentation of the case:* A 30-year-old mother delivered a male baby at 39 weeks through a normal vaginal delivery. Examination revealed multiple congenital anomalies in the form of an Omphalocele, extrophied bladder, imperforate anus, ambiguous genitalia and a large pelvic diastasis. Fecal matter was noted at the most inferior point of the extrophied bladder, raising the suspicion of a recto- vesical fistula.

An exploratory laparotomy showed a fistula between the rectum and the neck of the extrophied bladder. A sigmoid colostomy was carried out in addition to a mucous fistula. The fascial defect of the Omphalocele was approximated to the upper border of the extrophied bladder.

At the age of 2 years, the baby underwent a bladder extrophy repair, a posterior sagittal anorectoplasty and bilateral osteotomies.

*Discussion:* OEIS complex has been reported to occur with a wide variety of associated anomalies, and this necessitates a thorough investigation in order to formulate an appropriate treatment plan. A prenatal diagnosis of OEIS complex can be made by ultrasound stressing the importance of antenatal follow up and a multidisciplinary approach in management.

*Conclusion:* We described a rare variant of an OEIS complex and management of such anomalies requires a multidisciplinary input.

## 1. Introduction

Omphalocele, bladder extrophy, imperforate anus and spinal defect (known as OEIS) is a very rare congenital anomaly. The OEIS variant has an incidence of 1 in 200,000 to 400,000 live births [1,2]. The etiology is unknown and the occurrence of this anomaly is sporadic in nature [3].

In this report we describe a case of an OEIS variant associated with a wide pubic diastasis, bladder extrophy, recto bladder neck fistula and a high ano-rectal malformation. This work has been reported in line with the SCARE 2020 criteria [4].

## 2. Case presentation

A 30-year-old mother delivered a 3.28 Kg male baby at 39 weeks through a normal vaginal delivery. The parents have history of consanguinity. APGAR scores at 1 and 5 min were 8 and 9, respectively. Upon further assessment the baby was found to have multiple congenital anomalies in the form of an Omphalocele, extrophied bladder, imperforate anus, ambiguous genitalia and a large pelvic diastasis. A detailed assessment of the genitalia showed a single hemi phallus on the left side, 2 hemiscrota and bilateral impalpable undescended testes (Fig. 1). Fecal matter was noted at the most inferior point of the extrophied bladder, raising the suspicion of a recto-vesical fistula (Fig. 2).

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The baby was further assessed for a VACTERL association, and an ultrasound of the abdomen and pelvis revealed a bilateral grade I hydronephrosis, while an echocardiogram showed a patent foramen ovale. An ultrasound of the head was unremarkable. An ultrasound of the spine showed a normal vertebrae and a spinal cord terminating at the level of L2. An invertogram was performed and it raised the suspicion for high anorectal malformation. There was neither tracheo-esophageal anomalies nor limb defects.

A decision was made to operate at day 2 of life. An exploratory laparotomy was carried out, revealing the presence of the stomach, small bowel, and colon up to the level of the rectum. A fistula was noted between the rectum and the neck of the extrophied bladder. A bilateral ureter scope was unremarkable and ureteric stents were inserted. A sigmoid colostomy was carried out in addition to mucous fistula. The fascial defect of the Omphalocele was approximated to the upper border of the extrophied bladder. A multidisciplinary team was formed including nursing stuff, pediatric general surgeon, pediatric urologist, pediatric orthopedic, pediatric anesthetist, and neonatalologist in order to procedure with surgical and medical management of this complex pathology.

The postoperative course was uneventful. After a multidisciplinary team assessment, a decision was made to delay the closure of the extrophied bladder. At the age of 2 years, the baby underwent a bladder extrophy repair, a posterior sagittal ano-recto-plasty and bilateral osteotomies. Colostomy closure was carried out 3 months later. The patient had bladder continence and had no urinary tract infection. The postoperative course was uneventful.

Upon outpatient follow-up the baby was doing well. He had been walking without any walking aids, and he was developmentally equivalent to his peers. Neurological and musculoskeletal exam revealed no deficits. Further reconstructive surgery to the phallus was planned for a later stage.

## 3. Discussion

In 1978, Carey et al. reported the first case of OEIS complex, and the severity of this congenital multi- systemic malformation is variable and ranges from epispadias to bladder extrophy to cloacal extrophy [1]. We

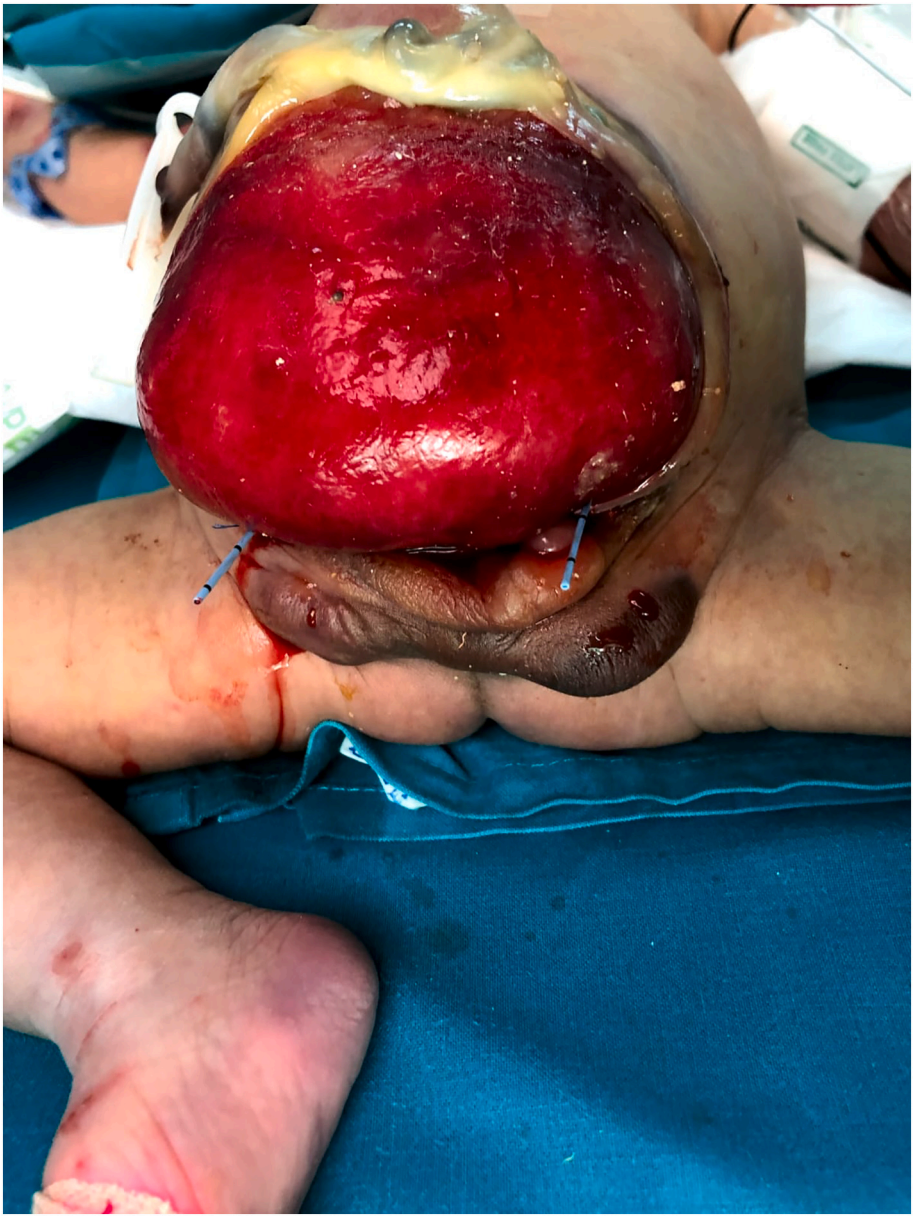

**Fig. 1.** This picture shows the orifice of the recto- extrophied bladder neck fistula with meconium stain.

present an unusual case of an OEIS variant associated with a wide pubic diastasis, bladder extrophy with a recto-bladder neck fistula and a high ano-rectal malformation.

Although the exact cause of OIES complex is uncertain and it's occurrence is sporadic, in some cases it has been associated with chromosomal aberrations e.g. deletion of chromosome 1p36 and homeobox gene [5–7,12]. Furthermore some cases have been reported to occur in association with maternal exposure to diazepam, smoking, maternal obesity and uterine fibroids [7]. Lubala TK et al. reported a case of OEIS complex correlated to maternal gonococcal infection in early pregnancy and fetal alcohol exposure throughout the pregnancy [8]. None the above risk factors were present in our case.

In addition to the cardinal malformations, OEIS complex has been reported to occur with a wide variety of associated anomalies. These include anomalies of the central nervous system, cardiovascular system, vertebrae, upper urinary tract, vertebral column and lower limbs. An absent appendix and malrotation have also been reported [9,10]. As a result we investigated for the occurrence of the wide array of associated anomalies.

A prenatal diagnosis of OEIS complex can be made by ultrasound as early as 16 weeks of gestation [11]. Despite the availability of major and minor radiological criteria that can aid in reaching a prenatal diagnosis, the full extent of anomalies cannot always be identified prenatally [11,12]. This patient had multiple antenatal ultrasound scans and none of them suspected the diagnosis of OEIS complex. We stress the importance of managing such anomalies through a multidisciplinary team approach as well as the importance of antenatal follow up.

Treatment upon diagnosis consists of immediately after birth the newborn is stabilized and the exposed organs should be protected by enclosing them in a bowel bag or by first moistening surfaces with saline and covering with sterile plastic wrapping [12]. Then the urologic exam is performed to know the genetic sex, size of hemi-bladders, and presence of spinal dysraphism and if the gender has not been previously determined or is not obvious on exam, Karyotyping can be performed [13]. In addition to the urologic evaluation, consultation with general surgery is required. Once the initial evaluation is complete, the parental

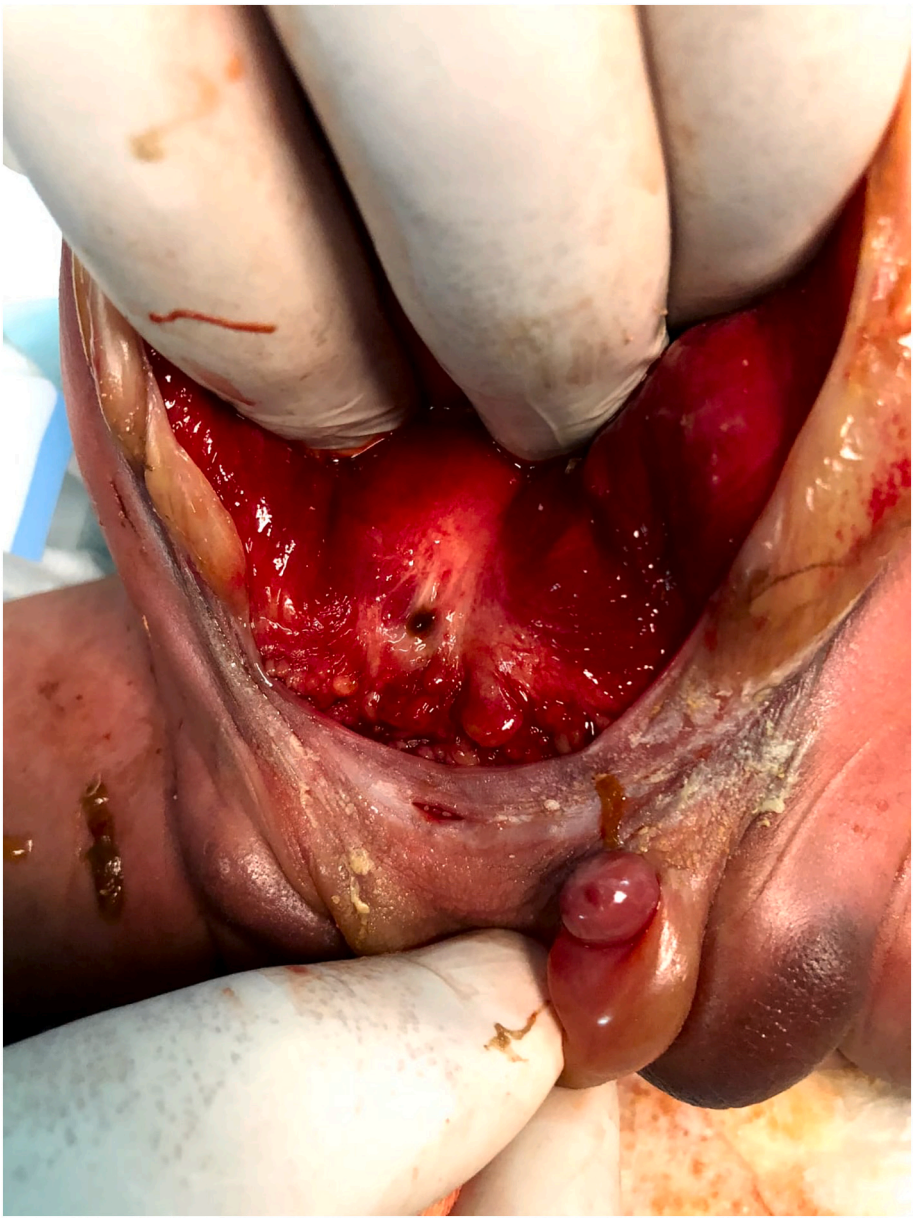

**Fig. 2.** This picture demonstrate the omphalocel and bladder exstrophy and both ureteric stents that were inserted. In addition to the above it clearly shows hemiphalus and hemiscrotum.

discussion is carried out concerning gender assignment, surgical reconstruction, possible functional deficits, and overall expected quality of life [13]. The aim of the management is a successful bladder closure, and the repair of the extrophied bladder is a very challenging pediatric surgery. The repair is either performed early within the first 2 to 3 days of age or delayed within 6 to 12 weeks of age. In our case the delayed repair is done.

The post-operative care provide a challenge of the medical team and parents. The patient must be attentively monitored regarding his nutrition, and initial use of total parental nutrition is supported. Moreover, the patient must be immobilized, and a broad spectrum antibiotics are advised as these patients have several drainage tubes [14]. A pediatric pain service is encouraged in order to control the pain. Finally, it is very important to limit any abdominal distension for a successful abdominal closure [15].

## 4. Conclusion

In this report we present a rare variant of OEIS complex, which is an unusual case of bladder exstrophy with rectobladder neck fistula. The incidence of this variant has not yet been established as there is not enough data in the literature. Management of such anomalies requires a multidisciplinary input, and once the initial evaluation is complete, a parental discussion should be made concerning the gender assignment, treatment plan, possible functional deficits, and overall expected quality of life.

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## Consent

Written informed consent was obtained from the patient relatives for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Ethical approval

This report is exempt from ethical approval (case report and literature review).

## Guarantor

Amar Alnaqi- MD, FRCSC. Dralnaqi@gmail.com

## Informed consent

Informed consent was obtained from the parents for publication of the case report and associated images anonymously.

## Registration of research studies

NA.

## CRediT authorship contribution statement

Athari AlShammari- data collection, literature review, writing.
Waleed Burhamah- writing, literature review, editing.
Amar Alnaqi- Supervision, review and editing.

## Declaration of competing interest

The authors declare no conflict of interest.

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