

Omphalocele, exstrophy of cloaca, imperforate anus, and spinal defect complex, multiple major reconstructive surgeries needed

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Abstract OEIS complex is a rare combination of serious birth defects including omphalocele, exstrophy of cloaca, imperforate anus, and spinal defects. The aim of managements has shifted from merely providing survival to improve patient outcomes and quality of life with higher level of physical and social independence. Multiple complicated reconstructive surgeries always needed for achieving the goals of treatment. In this case report, we aimed to present our surgical approach for this rare abnormality to achieve functionally and socially acceptable outcome.

Keywords: Cloacal exstrophy, imperforate anus, omphalocele, spinal defect, surgical repair

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Received: 23.12.2017, **Accepted:** 23.12.2017

INTRODUCTION

Carey *et al.* in 1978 was the first to use the term OEIS complex.^[1] It includes omphalocele, exstrophy of cloaca, imperforate anus, and spinal defect. The etiology remains unknown. Despite the sporadic nature of OIES complex it is thought to be multi-factorial including environmental exposures, twin presentation and *in vitro* fertilization.^[2,3] The omphalocele, exstrophy of cloaca, imperforate anus, and spinal defect (OEIS) complex may be associated with several anomalies (for example, genital abnormalities, renal malformations, symphysis pubis diastasis, and limb abnormalities) in addition to the main component.^[4]

Surgical repair is always challenging and requires that multidisciplinary teams include neonatologists, pediatric surgeon, pediatric urologist, pediatric orthopedist, pediatric

neurosurgeons, and genetic and pediatric endocrinologist. The surgical repair includes many surgeries at different ages with potential complications such as urogenital and gastrointestinal dysfunction, neurologic disorders, and psychological consequences.^[5] The prognosis for OEIS complex depends on the spectrum of the severity of the structural defects and the experience of the managing team.


CASE REPORT

Our male patient who is now 10 years old was delivered by full-term cesarean section due to twin presentation to nonconsanguineous couple with birth weight of 2.5 kg and a height of 45 cm and stable vital signs with a normal Apgar score. Antenatal history shows normal pregnancy with unremarkable history and normal ultrasound. Our

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How to cite this article: Neel N, Tarabay MS. Omphalocele, exstrophy of cloaca, imperforate anus, and spinal defect complex, multiple major reconstructive surgeries needed. *Urol Ann* 2018;10:118-21.

Access this article online	
Quick Response Code:	Website: www.urologyannals.com
	DOI: 10.4103/UA.UA_193_17

patient is the fifth child of the family – four brothers and one sister – who are all in healthy condition including his twin and nothing specific was noted on the other siblings.

The patient was delivered by a cesarean section due to twin presentation (first twin boy, one placenta) and was admitted to a private hospital due to congenital urogenital malformation and was transferred to our hospital at patient age of 15 days.

The patient was admitted in the neonatal Intensive Care Unit under a multidisciplinary team comprising neonatologist, pediatric surgeon, and pediatric urologist who were involved in the management, and also pediatric orthopedist, pediatric neurosurgeon, and genetic and pediatric endocrinologist were consulted and their opinion was shared in the management.

On examination, the baby had omphalocele covered with granulation tissue and foreshortened hindgut between two bladder hemiplates with ureteric orifice obvious in each hemiplate. Partially prolapsed terminal ileum and cecal surface were evident. There was wide separation between symphysis pubis and absent anal verge. Right and left hemiglans and haemophilus attached to corresponding pubic bones. Right and left scrotal compartment with left testis is palpable and the right one is not [Figure 1 and Table 1]. The karyotype of the baby was 46 xy.

After stabilization of the baby general condition, laboratory and radiological investigation was completed, and board meeting including the managing team was performed where results of radiology were discussed.

The left kidney was ectopic in the pelvic region with no hydronephrosis; however, the right kidney was normal in

position with grade 4 hydronephrosis. A nuclear study revealed nonobstructed right kidney with split function 19% and left kidney was 81%. Magnetic resonance imaging showed wide separation of the symphysis associated with spinal dysraphism and absence of the lower sacral segments with butterfly vertebra and hemivertebra in the thoracic region.

Neurosurgery team decided to observe the baby with no immediate surgical intervention at the same time pediatric orthopedic team was willing to share with bilateral iliac osteotomies.

After putting, the plan for surgery meeting with the parents was performed with explanation of the steps of surgery and expected outcomes and complications, informed consent was obtained, and the baby went for surgery.

The surgery was started by bilateral anterior iliac osteotomies then excision of the omphalocele membrane then separation of the cecal plate from the two hemibladders and tubularization, and an end colostomy was created from the distal colon in the left lower quadrant of the abdomen. The two bladder halves were sutured in the midline, and dissection from lateral skin attachments was performed then closure of the anterior wall of the bladder. However, in view of small bladder hemiplates, the bladder was closed and the bladder neck was sutured to the perineal region [Figures 2 and 3]. The two hemiphallus and hemiglans were approximated and penis was reconstructed.

The baby kept immobilized for 6 weeks and postoperative period went smoothly with no major complications.

At the age of 1 year, bilateral hernia repair with right orchidopexy was carried out.

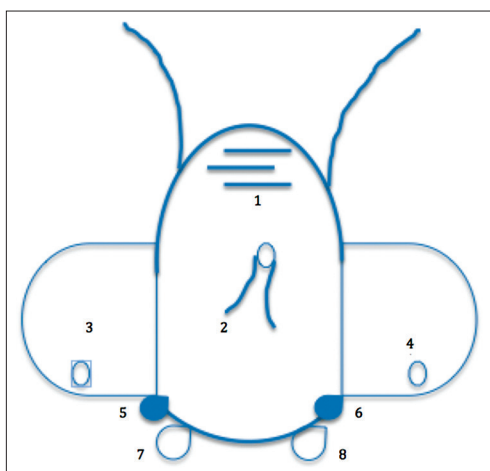


Figure 1: Illustration of anatomy



Figure 2: Preoperation



Figure 3: Preoperation

He was followed up regularly with pediatric surgery and pediatric urology, and during follow-up, he developed two attacks of intestinal obstruction which managed conservatively and he suffered from many attacks of recurrent urinary tract infections with drop of the split function of the right kidney from 19% to 8%.

At the age of 9 years, the patient was evaluated for continence and the pediatric surgery team decides that terminal colostomy is the end solution for him as the child has nearly no pelvic floor muscle.

For urinary continence and treatment of the recurrent urinary tract infection, the patient underwent right nephrectomy and augmentation ileocystoplasty with closure of the bladder neck after it was dissected from the perineal skin with creation of continent cutaneous outlet through Monti technique using subserous extramural tunnel (Mansoura technique) [Figure 4] as a continent mechanism.

The patient tolerated the procedure very well and he is now continent for urine through clean intermittent catheterization every 4 h through the continent cutaneous outlet with no more urinary tract infection.

The patient is now waiting for puberty to complete his repair through phalloplasty as the last step in his repair.

The patient described in this report showed classic presentation of OEIS complex as described by Carey *et al.* in 1978 (omphalocele, cloacal exstrophy, imperforate anus, and spinal abnormalities).^[1]

OEIS complex is usually sporadic with no obvious etiology. An observation was made that 10%–30% of cases occur in twin pregnancies. It was suggested that OEIS complex



Figure 4: Postoperation

and monozygotic twinning are early blastogenesis defect.^[3] Our case was twin presentation.

It is probably the most challenging birth defect in pediatric urology and surgery. Some considered that patient survival and optimal management of this anomaly reflect the maturity of pediatric surgery and urology and pediatric health care in general.^[6]

Over the past 30 years, the focus has shifted from the survival, to improving patient outcomes and ensuring the optimum quality of life.^[6]

The surgical treatment is difficult due to the complexity of these malformations, and the patients should be managed by multidisciplinary team.^[5]

In our case, multidisciplinary team included neonatologists, pediatric surgeon, and pediatric urologist who were involved in the management and also pediatric orthopedist, pediatric neurosurgeons, and genetic and pediatric endocrinologist were consulted and their opinion shared in the management.

Gender assignment is an important and debatable issue; historically, a genetically male infant with a phallus of inadequate size for reconstruction was often assigned to female gender, performing early orchidectomy with subsequent hormone replacement at puberty.^[6] The studies reported unclear conclusions on the possible psychosocial and behavioral outcomes of gender reassignment of genotypic males being raised as females.^[7] At the same time, advancement in the surgical techniques for phallic reconstruction, many are advocating for assigning gender according to karyotype.^[8]

In our case, the baby was raised as male according to the karyotype and no gender reassignment was offered.

Table 1: Figure 1 details

Number	Anatomy
1	Omphalocele covered with granulation tissue
2	Ileum partially prolapsed and nonobstructed
3,4	Right and left bladder hemiplate and ureteric orifice
5,6	Right and left hemiglans and haemophilus
7,8	Right and left scrotal compartment

The first surgery for correction of OEIS can be achieved in single or staged procedures.^[5]

In our case, single-stage procedure with bilateral anterior iliac osteotomies as the patient referred to us at the age of 15 days, and we proceeded for surgery at 20 days of his age, so approximation of the severely separated symphysis was mandatory by iliac osteotomy and postoperative immobilization.

In the surgery for augmentation, we tried to avoid excessive resection of the ileum to avoid short bowel syndrome, and we used Monti technique for creation of the continent cutaneous outlet using subserous extramural tunnel technique (Al Mansoura technique)^[9] as a continent mechanism.

The objectives of management include secure abdominal wall and bladder closure, preservation of renal function, prevention of short bowel syndrome, reconstruction of genitalia adequate functionally and cosmetically, and achievement of acceptable urinary and fecal continence.^[10]

In our case, we achieved most of the goals of management where we close the abdominal and bladder wall successfully and the renal function is maintained with no short bowel syndrome and the patient is currently continent for urine through clean intermittent catheterization through a continent stoma and we are waiting for puberty to go for phalloplasty which considered the last step of repair to achieve complete functional and social acceptable outcome.

The patient has stoma for colostomy which will be permanent as the pediatric surgery team found that there is no enough muscles in the perineal region enable them for creation of sphincter for stool continence.

In the past, it has drastic negative impacts on the full range of patient's genital and urinary function. However, advances in medical and surgical management have allowed for dramatically improved survival and continence rates, but even with the best care, these children will require lifelong support.^[6]

We present this case to show that with new surgical techniques and advances in medical care, we were able to

reach with our patient to the most acceptable functional and social outcome which helps him to integrate easily with the normal lifestyle.

CONCLUSION

The functional and social management goals for this rare congenital anomaly can be reached with multidisciplinary team through multiple complicated surgical procedures with very satisfying result.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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