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Case Report

# Clinical Case of Cloacal Exstrophy with OEIS Complex in a Premature Infant

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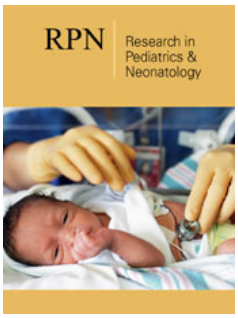


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## Abstract

The term "OEIS-complex" was introduced by Carey and colleagues to describe a group of malformations associated with cloacal exstrophy. These include: O - omphalocele, E - exstrophy, I - imperforate anus, and S-spinal deformities. Cloacal exstrophy, including the OEIS-complex, is typically diagnosed prenatally through modern imaging techniques, particularly ultrasound during the screening stage in antenatal care settings. This combined malformation generally has satisfactory treatment outcomes but requires multiple surgical interventions aimed at reconstructive surgery and restoring organ function. However, the long-term quality of life for patients is often compromised by severe disability, fecal incontinence due to functional disorders of the operated colon, and cognitive impairments. The extended duration of treatment, frequent surgical procedures, and the need for prolonged respiratory support in premature infants increase the risk of aspiration pneumonia, which is challenging to manage and can lead to sepsis and patient mortality. This article presents a clinical case of cloacal exstrophy in a premature infant complicated by aspiration pneumonia and sepsis.

**Keywords:** Cloacal exstrophy;OEIS -complex; Aspiration pneumonia; Sepsis

**Abbreviations:** OEIS: Omphalocele Exstrophy Imperforate Anus Spinal Deformities; CNS: Central Nervous System; SCT: Spiral Computed Tomography; MRI: Magnetic Resonance Imaging; SWI: Susceptibility Weighted Imaging

## Introduction

Cloacal exstrophy is a rare congenital malformation that encompasses the components of the OEIS-complex. The etiology of this intrauterine developmental defect remains incompletely understood but is currently considered multifactorial in nature, driven by both genetic and environmental factors. Clinical manifestations include the features of the OEIS-complex, as well as associated pathologies affecting the nervous, cardiovascular, and respiratory systems. Premature infants with cloacal exstrophy exhibit a heightened susceptibility to major clinical complications due to their prematurity. The primary approach to treating this condition involves a multidisciplinary effort by surgeons and neonatologists. Treatment includes sequential surgical interventions aimed at closing anatomical defects and pharmacological support focused on infection prevention and adequate pain management. The outcomes of comprehensive treatment are generally satisfactory and closely correlate with the severity of the congenital anomalies. However, long-term complications may arise, including impairments in urination, defecation, sexual function, and cognitive abilities.

## Case Presentation

Patient K. was born from the first complicated pregnancy and delivery, as the first of twins, at 36 weeks of gestation (prematurity). Birth weight: 1860g. Apgar score: 5/7. Multiple developmental anomalies were identified prenatally during routine ultrasound screening; however, the parents declined the option of artificial pregnancy termination.

The newborn received respiratory support (without continuous oxygen supplementation via nasal cannulas, with oxygen saturation maintained at 85%-86%) and thermal support and was started on artificial feeding from birth. On the second day of life, the infant was transferred from the neonatal intensive care unit to the neonatology department for further evaluation. A comprehensive examination was conducted, and multiple consultations with specialists were organized in a multidisciplinary format. A geneticist diagnosed sporadic OEIS-complex in the family with multifactorial inheritance. Ophthalmoscopy performed by a pediatric ophthalmologist revealed that the fundus was within the age-appropriate range. A pediatric urologist's consultation identified cloacal exstrophy, omphalocele, agenesis of the left kidney, and right-sided megaureter. Status localis: the mucous membrane of the cloacal exstrophy showed contact bleeding with isolated ulcerated areas. An endocrinologist diagnosed hypothyroidism, and hormone therapy with L-thyroxine was initiated at a dose of 12.5mcg 30-40 minutes before morning feeding, with the dose reduced to 6.25mcg after two weeks. A neurologist and neurosurgeon diagnosed hypoxic-hemorrhagic CNS damage, muscular hypotonia, cerebral depression syndrome, convulsive syndrome, and spina bifida in the lumbar spine.

During the SCT, the examination revealed a defect in the lower part of the anterior abdominal wall, through which loops of the intestine protrude beyond the abdominal cavity, along with a fragment of the anterior-lower edge of the liver. In the upper parts of the abdominal wall defect, the umbilical cord, measuring 56x85x97mm, is traced. Within the visible regions of the thoracic cavity, a heterogeneous soft tissue structure is identified para-aortally on the right, which requires dynamic observation. The right kidney is enlarged, with its parenchyma thinned to 3mm, reduced cortico-medullary differentiation, and a significantly expanded calyx-pelvic system. The ureter is tortuous and dilated up to 6 mm. Its distal part transitions into a rounded structure filled with contrast in the excretory phase, measuring 19x14x11mm, located at the entrance to the omphalocele at the level of the pelvis, which may correspond to a hypoplastic bladder. The excretory function of the right kidney is impaired. The left kidney is not reliably identified. The intestinal loops are pneumatized, with significantly reduced differentiation. The pelvic organs are not visualized. Additionally, there are anomalies in pelvic development: non-union of the pubic bones, retroversion of the iliac fossa, hypoplasia of the sacrum, and agenesis of the coccyx. Non-union of the posterior arches from L3 to L5, S1 to S2 is noted, as well as butterfly-shaped hemivertebrae at Th8-Th9.

During the MRI study, several abnormalities were detected. There were extraaxial subacute hemorrhagic layers in the left parietal region, measuring up to 2-3mm in thickness. SWI images revealed petechial hemosiderin inclusions in the cerebellar hemispheres and in the vascular plexuses of the lateral ventricles. There was also moderate edema of the soft tissues in the parietal-occipital region. Multiple vertebral malformations were observed, including butterfly-shaped hemivertebrae in the thoracic region, dysplastic vertebral bodies in the sacrum, and agenesis of the coccyx. Additionally, there was widespread hyperplasia of fatty tissue in the paravertebral and sacral regions, forming a lipomeningomyelocele with approximate dimensions of 20x36mm. At the level of L4, within the myelomeningocele, a possible splitting of the spinal cord elements resembling diastematomyelia could not be excluded. An extensive defect of the anterior abdominal wall was visualized, extending from the supra- and peri-umbilical area caudally, causing the prolapse of abdominal cavity structures outside. In the lower part of the protrusion, moderate free fluid was present, with visible, numerous tortuous mesenteric and peritoneal vessels. The umbilical cord was traced along the upper-lateral contour on the left. On the posterior left contour, a separate soft tissue component, measuring up to 12x19x10mm, was identified, which may correspond to hypertrophied and swollen labia or clitoris. The anus was not located in its typical position, and cicatricial-fibrous tissue was observed in its area. The rectum opened in the lower parts of the protrusion as a fistula. The right kidney was enlarged, measuring up to 27x32x55mm, with significant dilation of the calyx-pelvic system, while the left kidney was absent. A right-sided megaureter was present, with the ureter being tortuous. In its lower sections, it thinned to 2mm and lacked clear differentiation of the lumen. Near the distal portion of the right ureter, uterine tissue from a separated horn connected to the cervix was observed. The appendages were not visualized.

At this stage, polyantibiotic therapy was administered, including ampicillin-sulbactam, meropenem, ciprofloxacin, colistin, and tigecycline. Antifungal treatment consisted of a ten-day course of fluconazole. Hemostatic therapy involved tranexamic acid, phytomenadione, and blood transfusions, while symptomatic therapy included L-thyroxine and folic acid.

After partial stabilization of the child's condition by the sixth month of life, the decision was made to transfer her to the surgical department for the placement of an ileostomy to separate the digestive and urinary systems, preventing potential infectious complications.

Following the surgical procedure, the child was transferred to the pediatric department for further examination and treatment of a concurrent respiratory disease. A bacteriological culture was taken from the throat to isolate the pathogen and assess its sensitivity to chemotherapeutic agents. The cultures revealed Klebsiella pneumoniae and Acinetobacter baumanii, both of which were extremely multiresistant, resistant to all beta-lactams, including carbapenems. The procalcitonin level showed a dynamic increase, surpassing the normal range by 70 times, indicating the development of severe sepsis.

During the X-ray examination of the chest organs, a homogeneous darkening with clear contours was observed in the upper lung field, with an enriched pulmonary pattern on both sides. Inhomogeneous darkening of an infiltrative nature was detected in the upper and middle lung fields on both sides. The conclusion indicated atelectasis of the upper right lung and aspiration pneumonia.

The latest antibiotic therapy was initiated after the failure of the previous regimen, including colistin, cefotaxime, amikacin, and metronidazole. Symptomatic therapy involved omeprazole and paracetamol. Parenteral albumin was administered to address hypoproteinemia caused by multiple organ failure.

Despite extensive diagnostic, therapeutic, and surgical interventions, the child unfortunately passed away due to severe sepsis, septicemia, and ultimately, infectious-toxic shock and multiple organ failure [1-5].

## Conclusion

Cloacal exstrophy remains a complex congenital condition that requires a multidisciplinary approach to diagnosis, treatment, and rehabilitation. Timely prenatal diagnosis using modern imaging techniques, such as ultrasound and magnetic resonance imaging, is crucial for initiating early therapy and improving prognosis. Early surgical correction, combined with an appropriate postoperative therapeutic strategy, helps prevent severe acute complications, such as infections or sepsis, and reduces the risk of chronic dysfunctions in the genitourinary, digestive, nervous, and cognitive systems.

In clinical practice, cases like the one described in the article indicate a high risk of aspiration pneumonia and sepsis, which further increase the risk of mortality. This is particularly concerning in the context of prematurity, prolonged mechanical respiratory support, functional gastrointestinal disorders in children, and other aggravating factors related to medical care and the anatomical and physiological characteristics of both premature and full-term infants.

A coordinated effort from a team of specialists, dynamic monitoring of the patient's condition, and timely identification of complications are critical to successful treatment and minimizing risks to the lives of children with this pathology. Such an approach is essential for enhancing the quality of life for patients and their families.

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