

arches, narrow thorax, hydrocephalus, single umbilical artery)⁶.

According to the literature, sonographic findings of OEIS complex have been documented, but only few cases of prenatal diagnosis have been reported. Nevertheless, nowadays, the prenatal diagnosis of OEIS is possible by the identification of:

- A midline infra-umbilical defect with an irregular mass: in the inferior abdominal wall or cystic anterior wall structure (persistent cloacal membrane) or with omphalocele. Prenatal detection of anterior abdominal wall defect has significantly improved over last few years. It is among the more definitive diagnosis that can be made in a routine obstetric ultrasound examination. The Standard views required for prenatal ultrasound examinations, as per the guidelines of American College of Radiology, are the demonstrations of the umbilical cord insertion over foetal abdomen and the integrity of the anterior abdominal wall¹².
- Absence of the bladder between the two umbilical arteries.
- Lumbo-sacral myelomeningocele: always seen. Common spinal defect include hemivertebrae, sacral anomalies and either tethered cord and meningocele. But it is important to know that Källén et al wrote that the spinal defect may occur more cranially and are not restricted to the lumbo-sacral region⁸. Our case had caudal regression, there was abrupt disruption of vertebrae. The pathognomic feature is absence of vertebrae in axial section of the fetal abdomen.
- Anomalies of the inferior limbs are possible but club feet, limb duplication or amputations are generally not seen with OEIS¹⁰. Our case was a rare entity having club foot. (Figure 2)
- Wide pubic arch is classically present with symphysis pubis diastasis and congenital hip dislocation.
- Single umbilical artery: It is a frequent associated sign¹¹.
- Genital anomalies. Classically, the sex determination is often not possible. Other uro-genital anomalies are possible, including genital duplication. (Figure 1)
- Anal atresia. (Figure 2)
- Omphalocele. (Figure 3)

Majority of authors consider OEIS as a distinct syndrome, but there is a discussion if the exstrophy of the bladder sequence, exstrophy of the cloaca sequence or urorectal septum malformation sequence should be referred to distinct clinical entities.

Associated Anomalies: Different anomalies can be associated with OEIS complex:

- Cardiac anomalies: cardiac defects have been described with exstrophy of the cloaca alone (as atrial and ventricular defect).
- Renal anomalies
- Increased nuchal translucency: Only one case associated with OEIS was described by Schem et al². The most likely cause of increased nuchal translucency is probably vascular or hemodynamic, but the mechanism remains unclear. Nevertheless, there is no evidence for jugular lymphatic obstruction sequence in OEIS.

Markedly elevated serum levels of alfa fetoprotein is always present. But most cases of OEIS complex are diagnosed only at autopsy after interruption of pregnancy.

Prognosis: The prognosis of infants with OEIS complex is variable, depending on the severity of the structural defects. Survival will depend on the extension of the cloacalexstrophy and the neural tube defect. In less severe forms, good outcome with corrective surgery is possible but very uncommon. Cloacalexstrophy is lethal due to obstruction of the urinary tract and association with renal and pulmonary complications. So early prenatal diagnosis of OEIS complex is required to give parents the option to terminate the pregnancy. And it is also helpful to plan the appropriate perinatal management. In cases where parents decided to continue the pregnancy, serial scans are necessary to evaluate the progress of the ventriculomegaly, which can be associated with OEIS. But, we think that cesarean section should be performed in cases of OEIS complex to avoid dystocia and trauma.

Management: It seems essential to perform an early scan in the next pregnancy, because an accurate prenatal diagnosis of OEIS complex associated with malformations, is important for the detailed counseling of the family (interruption of pregnancy) and appropriate perinatal management by the obstetricians, pediatric surgeons, neurosurgeons, neonatologist, but especially anatomic-pathologist.

CONCLUSION

USG is the imaging modality of choice to diagnose OEIS Complex in the 2nd trimester. MRI can be useful back up to the USG findings.

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