Delayed recognition of bladder exstrophy and persistent cloaca: An Omphalocele exstrophy imperforate anus spinal abnormality (OEIS) Variant

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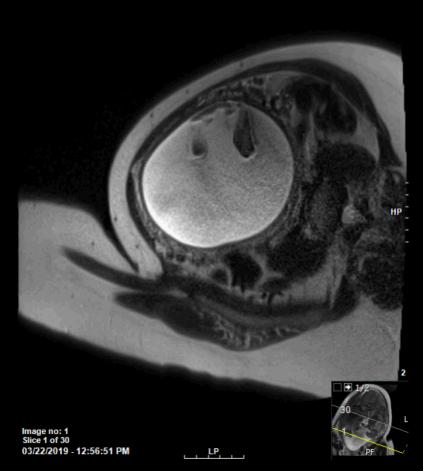




HPI

- Newborn term infant with abnormal prenatal ultrasound on 22- week anatomy scan.
 - Widened pubic diastasis
 - Dilated rectosigmoid colon
 - Mild polyhydramnios
 - Distended bladder in pelvis protruding into the omphalocele
 - Empties during exam
- Mother aged 31 G3P3, otherwise healthy
- Prenatal MRI obtained to confirm findings





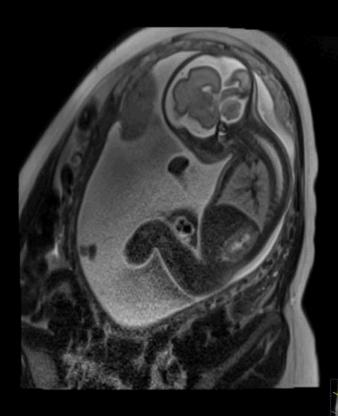
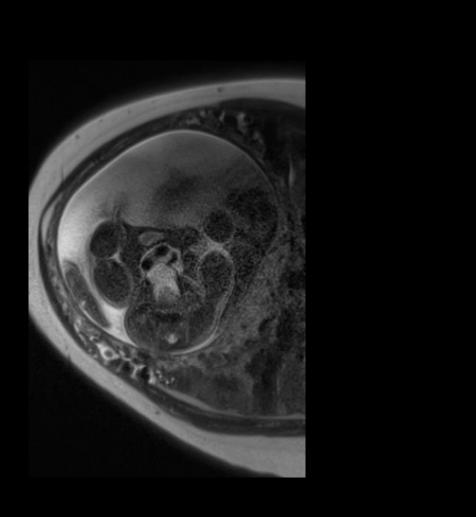


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Physical Exam

- Omphalocele
- Asymmetric labia/clitoral tissue
- Fistulous connection to the bladder
- Imperforate anus with a single mucosal-lined opening in perineum



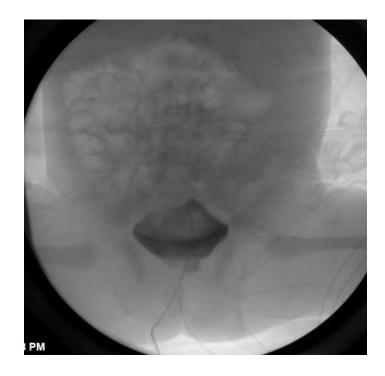
Diagnostic evaluation

- Cystoscopy/EUA
 - cloacal malformation
 - Appearance of an intact urethra/bladder neck complex
 - vaginal duplication
 - fistulous tract to the rectum

Diagnostic evaluation

- Cystogram
 - smooth-walled bladder without evidence of reflux

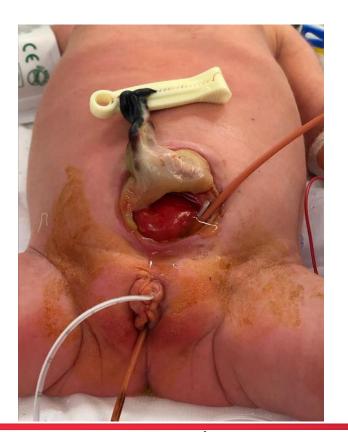
• Spinal ultrasound was normal.



DOL 4

Omphalocele rupture revealed bladder exstrophy





Hospital course

- She underwent a diverting colostomy on DOL 4.
- Discharged home with seran wrap coverage and prophylatic antibiotics
- Cloacal repair and closure of bladder exstrophy is planned at age 6 months of age.

Discussion

- Cloacal exstrophy is a rare and complex congenital anomaly
- Incidence of 1/200,000-400,000 live birth
- Occurs along a spectrum
- Associated omphalocele often contains bowel or liver
- Prenatal diagnosis of exstrophy is difficult to make. Often a diagnosis of omphalocele/gastroschisis made and the exstrophy overlooked.

Conclusion

 This case highlights the complexity associated with diagnosis of exstrophy- epispadias complex. Despite advances in prenatal imaging, diagnosis of this condition relies upon physical examination with the knowledge that each presentation can be different; not fitting in one box.

References

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- Carey J.C., Greenbaum B., and Hall B.D.: The OEIS complex (omphalocele, exstrophy, imperforate anus, spinal defects). Birth Defects Orig Artic Ser 1978; 14: pp. 253-263