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Case Report

Multidisciplinary management of VACTERL association in pregnancy: a cautionary tale

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ABSTRACT

VACTERL (vertebral defects, anal atresia, cardiac anomalies, tracheoesophageal fistula, renal anomalies, and limb abnormalities) association is a rare multisystem congenital disorder characterised by the presence of at least three of these characteristic anomalies. The study reports the case of a woman in her 30's (gravida 3 abortion 2) with VACTERL association of anal stenosis, patent ductus arteriosus and limb abnormalities presented in advanced labour at 38 weeks. Her antenatal course was uneventful, and she delivered a healthy neonate vaginally. On the 3rd postpartum day, she developed a wound infection. Despite antibiotic therapy and wound care, a small rectovaginal fistula persisted at six weeks, necessitating definitive surgical repair with perineal body reconstruction. The patient made a full recovery.

Keywords: Anal stenosis, Pregnancy, VACTERL, Rectovaginal fistula

INTRODUCTION

Vertebral defects, Anal atresia, Cardiac anomalies, TracheoEsophageal fistula, Renal malformations, and Limb abnormalities (VACTERL) is a rare, multisystem congenital disorder classically defined by the presence of at least three of these features.[1] Cardiac anomalies occur in 50-80% of these affected individual.[2] The diagnosis is established only after excluding other syndromic conditions with overlapping features, such as Coloboma, Heart defects, Atresia choanae, Retarded growth, Genital abnormalities, and Ear abnormalities (CHARGE) syndrome, Baller–Gerold syndrome, and DiGeorge syndrome. Unlike VACTERL syndrome, which implies a single defined genetic or embryonic cause, VACTERL association is not a true syndrome and no consistent gene, pathway or inheritance pattern is identified. A single unifying cause remains unfound, the most widely accepted pathophysiological mechanism attributes the condition to developmental field defects during blastogenesis affecting multiple organ system.[3] Advances in surgical techniques have greatly improved the prognosis and survival of the condition. Today, surgical corrections allow patients not only to achieve normal sexual and bowel function but reproductive function as well.

Consequently, obstetricians are increasingly encountering women with a history of VACTERL association during pregnancy and childbirth. Although clear guidelines regarding optimal route of delivery in women with prior anorectal malformation repairs are lacking, a systematic review of 13 attempted vaginal deliveries reported perineal tear in three women.[4] Here, we report the case of a woman with VACTERL association who had a vaginal delivery, highlighting the considerations for management in this unique clinical scenario.

CASE REPORT

A woman in her 30's (gravida 3, abortion 2), presented to the emergency department at 38 weeks of gestation in the second stage of labour. She has a known history of VACTERL association, diagnosed in infancy characterized by anal stenosis, patent ductus arteriosus (PDA), and limb defects (right thumb hypoplasia and left radial club hand). She underwent cutback anoplasty at 7 months of age for anal stenosis, followed by PDA ligation at 10 months. At 2 years, she underwent radialization of the ulnar bone of the left hand for radial club hand (Figure 1) and, at 10 years, pollicisation of the right index finger for congenital thumb hypoplasia (Figure 2). Other VACTERL-associated anomalies were excluded through appropriate screening during infancy. Following her childhood anoplasty, she experienced no long term defecatory dysfunction or bowel complications.



Figure 1: AP and lateral view of X-ray showing radialization of the hand.



Figure 2: AP and lateral view of X-ray showing pollicisation of the right index finger for congenital thumb hypoplasia.

She had been married for three years, with a history of two early pregnancy losses managed medically and surgically. Her antepartum period was uneventful, with normal baseline antenatal investigations. A detailed fetal anomaly scan performed in the second trimester was normal. She received routine care in the high-risk obstetric clinic, received aspirin 150 mg once daily until 36 weeks. Patient had regular cardiology consultation due to her cardiac condition. At 37 weeks a pelvic examination confirmed adequate pelvic dimensions and a digital rectal examination revealed normal sphincter tone. Ultrasound examination revealed a single live fetus in cephalic presentation with normal amniotic fluid volume, placenta was posterior and the estimated fetal weight was 2.6 kg. Given her history of perineal surgery, an elective caesarean section was planned. At 38 weeks of gestation she was presented in spontaneous labour and with crowning of the fetal head. On admission her pulse rate was 98/min, blood pressure was 117/78 mmHg, she had no pallor, her cardiovascular and respiratory system and neurological examination was unremarkable. Rapid labor progression precluded a cesarean delivery and she delivered a healthy 2.7 kg baby vaginally after an episiotomy which was sutured with absorbable suture material. A thorough neonatal evaluation on day 1 of life was normal.

On the 3rd postnatal day, she developed fever and pain at the perineal wound site. Examination revealed profuse purulent discharge, oedema, induration, and marked tenderness at the perineal suture site. Pus culture was obtained, and broad-spectrum intravenous antibiotics were started empirically. Due to persistent lack of clinical response after 48 hrs, the sutures were removed and it showed fecal staining of the wound, around 0.5 cm rectovaginal fistula noted, daily wound dressings initiated. The wound culture grew *Escherichia coli, Staphylococcus aureus,* and *Streptococcus dysgalactiae*; she was treated with intravenous amikacin and clindamycin for two weeks based on sensitivity reports. Plastic surgery consultation advised a delayed repair after six weeks, given her history of anoplasty. She was discharged on day 20 postpartum with instructions to perform a sitz bath daily, maintain perineal hygiene, and follow up every week.



Figure 3: Photograph of the vulva after reconstruction surgery at 6 weeks.

At six weeks, she reported improved faecal continence, with examination revealing a small (0.5 cm) fistulous opening at the 6 o'clock position, 1.5 cm from the vaginal introitus. Sphincter tone had returned to near-normal by the end of the puerperium. Surgical management included excision of scar tissue, isolation and inversion of the rectovaginal fistula tract, perineal body reconstruction, and vaginal mucosal T-band closure using absorbable sutures (Figure 3). Following secondary pelvic reconstruction, she

was followed up weekly for a month, then every three months. Pelvic floor physiotherapy was initiated 6 weeks following surgery. She is currently asymptomatic with a good quality of life and has no complaints on follow-up at one year after the surgical repair.

DISCUSSION

VACTERL association is a rare group of congenital anomalies presenting unique management challenges, with no single diagnostic test available.[5] While most literature has focused on managing these conditions in children, there is a noticeable gap in understanding the long-term outcomes for adults and the implications for reproduction. Thanks to advances in neonatal surgery and the development of multidisciplinary care teams, more individuals with corrected VACTERL association are reaching reproductive age, creating new obstetric challenges that need to be addressed.

Determining the optimal route of delivery in women with prior anorectal or pelvic floor reconstruction remains a subject of ongoing debate, largely due to paucity of high-level evidence-based data. Furthermore, a history of pelvic reconstruction increases the risk of obstetric sphincter injury during vaginal delivery. As a result, some authors advocate for elective cesarean sections in patients with substantial anorectal or spinal abnormalities.[6] There are similar case reports where epidural anesthesia was administered and the woman had a successful vaginal delivery when she presented in active labor.[7] A recent study on this matter concluded that caesarean section is preferable in patients with cloacal repair since the patients would have undergone extensive correction of the perineal body and vagina, it is assumed that these patients have increased risk of damage to the repaired structures during vaginal delivery as the scar tissue does not stretch.[4] Furthermore, Appiah et al emphasized the importance of screening for gynecological malformations with history of anorectal malformations as anatomical abnormalities affect the choice of delivery method.[8] Although patients are at risk for perineal injuries Iwai et al, Pena et al and Greenberg et al showed that a vaginal delivery is possible without complications in patients with anorectal malformations.[9-11] While Ljubic et al reported a vaginal delivery in a patient with an unrepaired cloacal dysgenesis type 2.[12] Studies on adult follow-up have shown that significant clinical issues can become apparent only after childhood, particularly when faced with the physiological stresses of pregnancy and childbirth.[13] Currently the decision to perform caesarean section is based on expert opinion, the severity of the condition and the experience of the obstetrician and the pediatric surgeon.[4] A thorough risk assessment must be performed considering the wellbeing of both the mother and the baby. Moreover, clinicians must weigh the risks of obstetric sphincter injury against the surgical risks of caesarean delivery, such as paralytic ileus and postpartum infection. Reports from affected women highlight the critical role of multidisciplinary planning during pregnancy.[14]

In our case, we highlight the obstetric consequences following prior anorectal reconstruction. Even when childhood repair yields satisfactory fecal continence, pelvic floor fibrosis and altered microvascular supply may remain. Under the mechanical strain of labor, these compromised tissues are highly susceptible to severe obstetric anal sphincter injuries (OASIS). The rectovaginal fistula observed in this patient likely reflect the vulnerability of surgically reconstructed tissue. Similar concerns have been raised in adult outcome studies, which demonstrate that the functional limitations of repaired anorectal malformations can emerge under mechanical strain.[3] A significant prevalence of vertebral anomalies can complicate regional anaesthesia. Anesthetic management also requires careful consideration.

In our case, the patient had undergone a cutback anoplasty for low anal stenosis which is a minor surgical procedure with minor muscle disruption unlike extensive pelvic reconstruction such as posterior sagittal anorectoplasty (PSARP) dense scar tissue and altered perineal anatomy routinely prompt elective Cesarean section.[9,10] Although a cesarean delivery was initially planned, advanced labour necessitated a vaginal birth, emphasizing the importance of proactive counselling and early hospital admission for high-risk pregnancies. The postpartum challenges in this case reflect the impaired healing capacity of scarred perineal tissue. Early suture removal, aggressive wound care, targeted antimicrobial therapy and secondary reconstruction in close collaboration with plastic surgery were key to achieving full functional restoration in our patient, showing the importance of comprehensive, multidisciplinary care in managing complex obstetric injuries among women with congenital pelvic anomalies. Although the VACTERL association is typically sporadic, careful antenatal screening for anomalies is essential due to a small but recognised risk of recurrence.[3] Adult outcome studies suggest that many individuals with VACTERL can achieve a high level of functional independence when managed appropriately, which encourages optimistic reproductive counselling and emphasizes the need for tailored obstetric planning.

CONCLUSION

This case contributes valuable insights to the limited literature on pregnancy in women with VACTERL association and underscores the possibility of successful pregnancy outcomes. Women with VACTERL association can achieve successful vaginal delivery; however, prior anorectal surgeries may increase the risk of perineal trauma and wound complications during the peripartum period. Hence, it is advisable to do an elective caesarean section for women with previous extensive pelvic floor surgeries. Careful shared decision-making during the time of labour, close postpartum monitoring, and timely multidisciplinary intervention are essential to ensure optimal outcomes.

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REFERENCES

1. Gregg AR, Beltran-Regalado BA, Montero ML, Panda RM, Cucalon Calderon J. Management of vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula or atresia, renal anomalies, and limb abnormalities (VACTERL) in a child with complex medical needs in the foster care system: a pediatric case study. Cureus. 2024;16(7):e65581.
2. Cunningham BK, Hadley D, Hannoush H, Meltzer AC, Niforatos N, Pineda-Alvarez D, et al. Analysis of cardiac anomalies in VACTERL association. Birth Defects Res A Clin Mol Teratol. 2013 Dec;97(12):792-7.
3. Raam MS, Pineda-Alvarez DE, Hadley DW, Solomon BD. Long-term outcomes of adults with features of VACTERL association. Eur J Med Genet. 2011;54(1):34-41.
4. de Waal AC, van Amstel T, Dekker JJML, Ket JCF, Kuijper CF, Salvatore CM, et al. Outcome after vaginal delivery of women with a previous medical history of surgically corrected anorectal malformations: a systematic review. BMC Pregnancy Childbirth. 2023 Feb 4;23:94.
5. Sun M, Zhao Q, Yang B, Liu L, Zhou C, Yao X, et al. Molecular mechanism, diagnosis, and treatment of VACTERL association. Front Pediatr. 2025;13:1609624.
6. Tonks KE, Birch S, Ito Y, Bhagwat A. Perioperative management of a parturient with VACTERL association for a caesarean section. Anaesth Rep. 2023;11(1):e12213.
7. Ramos JA, Shettar SS, James CF. Neuraxial analgesia in a parturient with the VACTERL association undergoing labor and vaginal delivery. Braz J Anesthesiol. 2018;68(2):205-8.
8. Appiah-Sakyi KK, Abdel-Aty M. Childbirth following previous repair of an imperforate anus. J Obstet Gynaecol. 2009;29(1):62-3.
9. Iwai N, Deguchi E, Kimura O, Kubota Y, Ono S, Shimadera S. Social quality of life for adult patients with anorectal malformations. J Pediatr Surg. 2007;42(2):313-7.
10. Peña A, Levitt MA, Hong A, Midulla P. Surgical management of cloacal malformations: a review of 339 patients. J Pediatr Surg. 2004;39(3):470-9.
11. Vaginal delivery after cloacal malformation repair. Obstet Gynecol. 1997. Available from: https://journals.lww.com/greenjournal/fulltext/00006250-199710001-00010~vaginal-delivery-after-cloacal-malformation-repair, Accessed on 5-April 2026
12. Ljubić A, Sulović V, Stanković A, Cvetković A. Cloacal dysgenesis and vaginal delivery [in French]. J Gynecol Obstet Biol Reprod (Paris). 1993;22(4):417-8.
13. Kassa AM, Engstrand Lilja H, Engvall G. From crisis to self-confidence and adaptation: Experiences of being a parent of a child with VACTERL association—a complex congenital malformation. PLoS One. 2019;14(4):e0215751.
14. Milhem MI, Al-Junaidi SZ, Jabareen AE, Housheya MZ, Khamayseh WM, Milhem MI. VACTERL association: a case report highlighting multisystem anomalies and the critical need for multidisciplinary care in Palestine. Int J Surg Case Rep. 2026;138(4):1283-6.

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