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CLINICAL INFORMATION

Neuraxial analgesia in a parturient with the VACTERL association undergoing labor and vaginal delivery



Juan A. Ramos^{a,*}, Shashank S. Shettar^b, Christopher F. James^{a,c}

^a Department of Anesthesiology, University of Florida Health, Gainesville, Florida, USA

^b Department of Anesthesiology, University of Florida Health-Jacksonville, Jacksonville, Florida, USA

^c Department of Anesthesiology, Mayo Clinic, Jacksonville, Florida, USA

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Abstract

Introduction: The term VACTERL is an acronym for an association of congenital malformations: including vertebral, anal, cardiac, tracheo-esophageal, renal and limb anomalies. VACTERL anomalies pose a formidable challenge to anesthesiologists. We describe the anesthetic management of a parturient with VACTERL association, who underwent neuraxial analgesia for labor and vaginal delivery.

Case report: A 23 year old primigravida at 39 weeks gestation presented in labor at 4 cm cervical dilatation, completely effaced, requesting labor analgesia. Past medical history included VACTERL association with an imperforate anus and a partial endocardial cushion defect, both repaired in early childhood. She also had significant dorso-lumbar scoliosis with an extra lumbar vertebra. An MRI performed at 14 years age revealed the above findings with no spinal cord abnormalities. With a normal neurologic exam, a combined spinal epidural technique was performed. Despite significant scoliosis, the epidural space was identified at approximately the L3–L4 interspace at a depth of 5 cm. Spinal Fentanyl 25 mcg was administered followed by continuous patient-controlled epidural analgesia. The patient experienced excellent pain relief throughout her labor, and had an uneventful vaginal delivery 5 h after epidural placement.

Discussion: The rarity of VACTERL association in the obstetric population with its extensive anomalies mandates a multidisciplinary approach in the prenatal period as it can pose major challenges to all health care providers, including airway, ventilatory, cardiac and neuraxial problems. This is the first reported case of a successful and safe neuraxial technique in a laboring patient with the VACTERL association with albeit limited vertebral and spinal cord anomalies.

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* Corresponding author.

E-mail: jramos@anest.ufl.edu (J.A. Ramos).

PALAVRAS-CHAVE

Síndrome de Vacterl;
Epidural analgesia;
Dor obstétrica

Analgesia neuraxial em uma parturiente com síndrome de Vacterl em trabalho de parto normal**Resumo**

Introdução: O termo Vacterl é um acrônimo para uma associação de malformações congênicas que inclui anomalias vertebral, anal, cardíaca, traqueal, esofágica, renal e dos membros (*Limbs* em inglês). As anomalias Vacterl representam um enorme desafio para os anesthesiologistas. Descrevemos o manejo anestésico de uma parturiente com síndrome de Vacterl submetida à analgesia neuraxial para parto normal.

Relato de caso: Paciente primípara, 23 anos de idade, 39 semanas de gestação, apresentado em trabalho de parto 4 cm de dilatação cervical, apagamento completo, exigindo analgesia de parto. A história médica incluía síndrome de Vacterl com ânus imperfurado e defeito parcial do coxim endocárdico, ambos corrigidos na primeira infância. A paciente também apresentava escoliose dorso-lombar acentuada com uma vértebra lombar adicional. Uma RM realizada aos 14 anos de idade revelou os achados mencionados acima sem anormalidades na medula espinhal. Com um exame neurológico normal, a técnica anestésica combinada raqui-peridural (CRP) foi realizada. Apesar de escoliose significativa, o espaço peridural foi identificado próximo ao interespaço L3-L4 a uma profundidade de 5 cm. Fentanyl (25 mcg) foi administrado por via espinhal, seguido de analgesia peridural contínua controlada pela paciente. A paciente sentiu grande alívio da dor durante todo o trabalho de parto; o parto vaginal ocorreu 5 horas após a anestesia CRP sem intercorrências.

Discussão: A raridade da síndrome de Vacterl na população obstétrica com suas extensas anomalias exige uma abordagem multidisciplinar no pré-natal porque pode representar grandes desafios para todos os prestadores de cuidados de saúde, incluindo problemas respiratórios, de ventilação, cardíacos e do neuroeixo. Este é o primeiro caso relatado de uma técnica neuraxial bem-sucedida e segura em uma paciente em trabalho de parto com síndrome de Vacterl, embora com anomalias limitadas da coluna vertebral e medula espinhal.

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Introduction

VACTERL association is a rare multiorgan congenital disorder, usually defined as the simultaneous presence of at least three of the following anomalies from its acronym: vertebral defects, anal atresia, cardiac defects, trachea-esophageal fistula, renal dysplasia and limb abnormalities, without clinical or laboratory evidence suggesting other differential diagnosis¹ (Table 1).¹⁻⁴ Though first described 40 years ago by Quan and Smith,⁵ a single unifying cause remains unfound; with the most accepted explanation relating the disorder to developmental field defects during blastogenesis which phenotypically affect multiple organ systems.^{1,6}

Advances in surgical techniques and specialized care have greatly improved prognosis and survival in VACTERL association infants,¹ making it more common for the modern day anesthesiologist to encounter these patients as adults in day-to-day clinical practice. We present a case of a successful neuraxial block in a laboring parturient with VACTERL association.

Case report

A 23 year-old primigravida with a height of 165 cm, weight 73.5 kg and a BMI of 27–39 week intra-uterine pregnancy presented to the labor and delivery suite in active labor

with an 8 out of 10 pain (based on a 10 point visual analog scale). Unfortunately, despite adequate obstetric prenatal care she was never referred for high-risk obstetric anesthesiology antenatal assessment for unknown reasons. Her obstetric exam on admission was 4 cm cervical dilatation, with complete cervical effacement and at –2 station. Anesthesiology was consulted for neuraxial analgesia. Upon interrogation the patient referred a past medical history of VACTERL association diagnosed as an infant, and she had not been previously seen by the anesthesia service. The VACTERL association included the following anomalies in this patient: thoraco-lumbar levo-scoliosis, extra lumbar vertebra, incomplete endocardial cushion defect and anal atresia. The anal atresia required a colostomy at birth and colostomy takedown by age 2 years, with subsequent procedures, which included multiple hernia repairs and scar revisions. At age 3 years, the patient underwent repair of her incomplete endocardial cushion defect, which included an atrial septal defect repair, repair of the transitional canal and suture repair of a cleft mitral valve. The patient denied any cardiac symptoms on admission. Her present cardiac exam included a Grade II–III/VI systolic ejection murmur heard at the left sternal border. Electrocardiogram revealed a normal sinus rhythm. An echocardiogram during this gestation and five months prior to admission revealed an ejection fraction of 65%, mild mitral and aortic regurgitation and mild left ventricular outflow obstruction.

Table 1 VACTERL association with its group of congenital anomalies.

Vertebral	Segmentation defects such as hemivertebrae, "butterfly vertebrae", "wedge vertebrae" (dysplastic vertebrae), vertebral fusions, supernumerary or absent vertebrae, and other forms of vertebral dysplasia.
Vascular	Single umbilical artery.
Ano-rectal	Imperforate anus/anal atresia.
Cardiac	Ventricular septal defect, atrial septal defects and tetralogy of Fallot. Less common defects are truncus arteriosus and transposition of the great arteries.
Tracheo-esophageal	Esophageal atresia with tracheo-esophageal fistula.
Renal	Unilateral renal agenesis (or bilateral in severe cases), horseshoe kidney, cystic and/or dysplastic kidneys.
Limb	Radial anomalies, thumb aplasia/hypoplasia and polydactyly.
Neurologic	Tethered spinal cord, syrinx, tight filum terminale and lipomeningomyelocele.
Miscellaneous	Ambiguous genitalia, abdominal wall defects, diaphragmatic hernia and oligohydramnios sequence defects.

Unfortunately, information concerning her vertebral anomalies was scant with no imaging studies of vertebral and/or spinal anatomy. However, documentation from the patient's pediatric orthopedic surgeon and an MRI report nine years earlier did not reveal any spinal cord manifestations or other vertebral issues. Airway exam revealed a Mallampati class II airway, 7 cm thyromental distance and intact cervical range of motion. The patient's laboratory values on admission included a hemoglobin of 12.5 g.dL⁻¹ and a platelet count of 274,000 μ L.

Despite limited data on her skeletal anatomy, except for her obvious scoliosis and an old orthopedic document, along with her lack of neurologic symptoms, and compounded by her severe labor pain, a neuraxial block for labor and vaginal delivery was deemed a reasonable option and the patient underwent combined spinal-epidural (CSE) analgesia. The patient's scoliotic anatomy was marked and using the inter-cristal landmark, a lower lumbar (L3-4 or 4-5) interspace was used. Excellent spinous processes and interspaces were palpated so ultrasound utilization was deferred based on the patient's imminent distress. The epidural space was identified at 5 cm with a saline loss-of-resistance technique using a 3.5 inch 17 gauge Tuohy needle, followed by a subarachnoid puncture with a 5-inch 25 gauge pencil-point Pencan[®] spinal needle with administration of 25 μ g of intrathecal fentanyl. Subsequently a 20 gauge closed tip catheter was threaded and secured at 9 cm at the skin. With prior negative aspiration a 0.1% bupivacaine and 2 μ g per mL of fentanyl epidural infusion was started with a patient controlled epidural analgesia (PCEA), consisting of a continuous infusion of 6 mL.h⁻¹ with a 5 mL bolus with a 20 min lock out. The patient experienced pain relief within 5 min of administering the spinal dose and went on to achieve a satisfactory T10 dermatome level. Five hours after the neuraxial block, the patient underwent an uneventful vaginal delivery of a viable male infant weighing 2.65 kg and Apgars of 9¹ and 9⁵. The epidural catheter was removed intact without any anesthetic complications.

Discussion

VACTERL association is a cluster of congenital malformations, which present together more frequently than

by chance. Its frequency is estimated to be between 1/10,000 and 1/40,000 infants⁷ with approximately 70% male preponderance,⁸ making it a rarity, especially within the obstetric population. A review of the literature yielded only two cases (a case report and a letter to the editor) in which anesthesia was provided for scheduled elective cesarean section in these two pregnant patients with VACTERL association.^{9,10} Epidural anesthesia was used for one case, and a combined general and an epidural anesthetic was used in the second case. In contrast, our case was the first one involving successful neuraxial analgesia (CSE) in a patient who presented in active labor with VACTERL association, without previous anesthetic consultation for labor and vaginal delivery. Several challenges and learning points can be ascertained from this experience and the available literature, especially considering the low number of high-risk obstetrical anesthesia candidates (<25%)¹¹ who are actually referred and assessed by an anesthesiologist in the prenatal phase.

Cardiac anomalies occur in 50–80% of these cases and severe cardiac anomalies were present in our case. However, repair of these cardiac anomalies occurred at age 3 years, fortunately, resulting in minor residual signs and symptoms that minimally restricted our patient's normal daily activities. Obviously, cardiac compromise would have compounded our anesthetic concerns in this case. Vertebral anomalies are a hallmark of VACTERL association, present in approximately 60–80% of patients. The range vertebral anomalies are broad, and include defects such as hemivertebrae, vertebral fusions, and supernumerary or absent vertebrae and dysplastic vertebrae such as "butterfly vertebrae", "wedge vertebrae".^{1,3} In some cases clinical scoliosis may be the first sign of vertebral anomalies,⁴ which should be investigated in depth with imaging and also lead to significant restrictive ventilatory defect. In addition, neurologic anomalies such as tethered spinal cord (TSC), syrinx, tight filum terminale, and lipomeningomyelocele have also been described in patients with colorectal and urogenital abnormalities in VACTERL association, usually with neurologic manifestations.^{3,4} In a small prospective study² seven out of nine (78%) infants with VACTERL association were found to have TSC by magnetic resonance. This highlights the importance of close neurologic/orthopedic follow-up of

these patients. Fortunately, despite the lack of prior consultation or knowledge of this patient in early gestation, along with the lack of imaging of her skeletal anomalies, the documentation by the patient's childhood orthopedic surgeon and the patient's denial of neurological signs and symptoms and her limited vertebral anomalies allowed for a successful neuraxial analgesic technique. However, in many cases of the VACTERL association, neuraxial techniques may be difficult or contraindicated secondary to severe scoliosis, obscure anatomy or more extensive vertebral anomalies, in which case ultrasound use for anatomic assessment is highly advocated. In the setting of severe restrictive lung disease, neuraxial analgesia may be a viable option for labor and vaginal delivery, however, neuraxial anesthesia for a cesarean section may further compound any respiratory compromise. Another anesthetic concern with these patients, which was not a factor in this case, is the potential for a difficult airway in the setting of more extensive skeletal anomalies, which would include scoliosis involving and including the cervical vertebrae.

This report demonstrates that neuraxial placement for analgesia for labor and vaginal delivery can be performed safely in select patients with VACTERL association, however it also strongly encourages thorough and timely work-up of these patients, preferably in the prenatal setting, to minimize the risk of potential complications from neuraxial and/or general anesthetic techniques.

Consent

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

Conflicts of interest

The authors declare no conflicts of interest.

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