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Case report

Anesthetic management of a schoolboy with uncorrected truncus arteriosus type I, and severe pulmonary hypertension undergoing repair of congenital dislocation of the knee. Case report☆

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ABSTRACT

Introduction: The presence of truncus arteriosus represents just 1.2–3% of all complex congenital heart pathologies and if not corrected, less than 20% survive beyond one year of life. When the disease progresses patients usually develop severe pulmonary hypertension and may even develop into Eisenmenger's syndrome. The paper discusses a case of a schoolboy with a diagnosis of uncorrected truncus arteriosus type I, and severe pulmonary arterial hypertension undergoing non-cardiac surgery.
Case discussion: This is a 9-year old schoolboy with complex heart disease and similar pulmonary and systemic blood pressures, undergoing elective orthopedic surgery under regional anesthesia, with lumbar plexus block and posterior sciatic block. This anesthetic approach provided adequate anesthesia with hemodynamic stability and no impact on vascular resistance.
Conclusion: The choice of the anesthetic technique should be a planned decision based on the cardiovascular pathophysiology of the truncus arteriosus, the level of pulmonary hypertension, and the surgical procedure to be performed. Patients with severe pulmonary hypertension are at increased risk of developing suprasystemic pulmonary pressures with

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considerable hemodynamic involvement. Consequently, the anesthetic technique chosen shall provide adequate anesthesia and ensure the least hemodynamic impact. Whenever possible, it is important to consider the peripheral nerve block as the first line approach for orthopedic surgery.

Manejo anestésico de un escolar con tronco arterioso tipo I no corregido e hipertensión pulmonar severa sometido a reparación de luxación congénita de rodilla. Reporte de caso

Palabras clave:
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RESUMEN

Introducción: La presencia de tronco arterioso representa únicamente del 1.2 al 3% de las cardiopatías congénitas complejas y de no ser corregida, menos del 20% sobreviven después del año de vida. Si la patología progresa usualmente desarrollan hipertensión arterial pulmonar severa y pueden manifestarse incluso como un Síndrome de Eisenmenger. Se presenta un caso de un escolar con diagnóstico de tronco arterioso tipo I no corregido e hipertensión arterial pulmonar severa llevado a cirugía no cardiaca.

Presentación del caso: Escolar de 9 años de edad con cardiopatía compleja y presión arterial pulmonar similar a la presión arterial sistémica sometido a cirugía ortopédica electiva bajo anestesia regional con bloqueo de plexo lumbar y bloqueo ciático posterior. Esta técnica anestésica nos proporcionó una adecuada anestesia con estabilidad hemodinámica sin repercusión en las resistencias vasculares.

Conclusión: La elección de la técnica anestésica debe ser planeada en base a la fisiopatología cardiovascular del tronco arterioso, del grado de hipertensión pulmonar y del procedimiento quirúrgico a realizarse. Los pacientes con hipertensión pulmonar severa tienen mayor riesgo de manifestar presiones pulmonares suprasistémicas con compromiso hemodinámico importante, por lo que la técnica anestésica elegida será aquella que produzca una adecuada anestesia y menor repercusión hemodinámica. Es importante considerar de ser posible, el bloqueo de nervios periféricos como primera elección en cirugía ortopédica.

Introduction

Congenital heart disease continues to be one of the primary anomalies at birth. About 30% of these patients may require some type of non-cardiac surgery during their first year of life and it is estimated that up to 80% grow up to become adults in the United States.[1] Unfortunately in our environment there is a large number of children with congenital heart disease. Late diagnosis and lack of treatment result in severe complications that significantly increase perioperative morbidity and mortality.

Truncus arteriosus represents only 1.2–3% of the complex congenital heart diseases. If uncorrected, less than 20% of these patients will survive their first year of life.[2] The development of pulmonary hypertension is a serious complication of this type of condition and is characterized by a mean pulmonary artery pressure above 25 mmHg at rest and above 30 mmHg during exercise.[3] Our objective is to discuss a case report of a schoolboy diagnosed with uncorrected truncus arteriosus and severe pulmonary hypertension undergoing an elective orthopedic procedure.

Case presentation

9-year old male patient with a diagnosis of uncorrected truncus arteriosus type I, and severe pulmonary hypertension that presents with severe right knee pain and gate limitation. The MRI shows intact cruciate ligaments and discoid lateral meniscus. The patient was evaluated at the service of Pediatric Orthopedics and diagnosed with congenital right knee dislocation and was programmed for surgical repair.

The pre-anesthesia evaluation reported uneventful family history with a diagnosis of velocardiofacial syndrome and mild psychomotor retardation. The findings of the physical evaluation indicated a cooperative patient, functional class NYHA II, generalized cyanosis, precordial fremitus, multifocal systolic murmur, pulse oximetry 87%, and body weight of 22 kg. The transthoracic echocardiographic examination confirmed the cardiology diagnosis (Image 1). The patient has a history of anesthetic sedation for cardiac catheterization showing that the pulmonary pressure equaled the systemic blood pressure.