Rev Esp Anestesiol Reanim. 2018;65(4):234–237

Revista Española de Anestesiología y Reanimación

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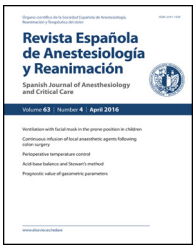

CASE REPORT

Cardiac arrest related to anaesthesia in Williams-Beuren syndrome☆

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Received 14 August 2017; accepted 13 November 2017
Available online 8 March 2018

KEYWORDS
Williams syndrome;
Cardiac arrest;
Anaesthesia;
Cardiopathy;
Death sudden;
Myocardial ischaemia

Abstract Williams-Beuren syndrome is the clinical manifestation of a congenital genetic disorder in the elastin gene, among others. There is a history of cardiac arrest refractory to resuscitation manoeuvres in anaesthesia. The incidence of myocardial ischaemia is high during anaesthetic induction, but there are patients who do not have this condition yet also have had very serious cardiac events, and issues that are still to be resolved. Case descriptions will enable the common pathophysiological factors to be defined, and decrease morbidity and mortality. We report the case of a 3-year-old boy with cardiac arrest at induction, rescued with circulatory assistance with extracorporeal membrane oxygenation and hypothermia induced for cerebral protection.

☆ Please cite this article as: Lucena Delgado J, Sanabria Carretero P, Durán la Fuente P, Gónzalez Rocafort A, Castro Parga L, Reinoso Barbero F. Parada cardíaca relacionada con la anestesia en el síndrome Williams-Beuren. Rev Esp Anestesiol Reanim. 2018;65:234–237.

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PALABRAS CLAVE
Síndrome Williams;
Parada cardiaca;
Anestesia;
Cardiopatía;
Muerte súbita;
Isquemia miocárdica

Parada cardíaca relacionada con la anestesia en el síndrome Williams-Beuren

Resumen El síndrome de Williams-Beuren es la manifestación clínica de una alteración genética congénita en el gen de la elastina, entre otros. Existen antecedentes de parada cardíaca refractaria a maniobras de resucitación en contexto anestésico. Es alta la incidencia de isquemia miocárdica durante la inducción anestésica, pero existen pacientes que, sin esta causa, también presentan eventos cardíacos muy graves. Quedan cuestiones aún por resolver. La descripción de casos permitirá definir factores fisiopatológicos comunes y disminuir la morbimortalidad. Presentamos el caso de un niño de 3 años con parada cardíaca en la inducción anestésica, rescatado con asistencia circulatoria con membrana de oxigenación extracorpórea e hipotermia inducida como protección cerebral.

Introduction

Cases of anaesthesia-induced cardiac arrest and death have been described in patients with Williams-Beuren syndrome (WS). Most of these are associated with coronary ischaemia. However, cases of acute coronary changes due to drug allergies have also been reported, showing that the triggering factor is not entirely clear[1] and there is growing interest in determining the origin of these high morbidity and mortality events. WS is the consequence of a genetic alteration that mainly involves the elastin gene.[2] It occurs in about 1/10,000 live births. The result is an array of multisystemic alterations (arterial stenosis, neuropsychological, urinary tract and gastrointestinal abnormalities, hypercalcaemia and diabetes mellitus, among others) with a characteristic phenotype (peculiar facies and mental retardation). During anaesthesia, these alterations can have significant cardiovascular repercussions. We present the case of a boy who presented cardiac arrest associated with anaesthesia induction.

Case report

A 3-year-old boy weighing 11.6 kg, with no known allergies, diagnosed with WS, scheduled for aortic arch enlargement due to severe supravalvular aortic stenosis (SVAS) and pulmonary artery stenosis. He had undergone enlargement angioplasty with a patch 2 years earlier. In the immediate postoperative period of that procedure he presented cardiac arrest, which was resolved with resuscitation for 10 min. He was currently under treatment with propranolol due to high blood pressure, and had non-specific chest pain with dyspnoea on moderate exertion, mild general hypotonia and psychomotor retardation of speech and gait. The electrocardiogram showed signs of left ventricular hypertrophy (VH) and repolarisation changes on lead III (Fig. 1). The cardiac ultrasound study showed mild pulmonary, aortic and mitral insufficiency, ejection fraction 48%, SVAS gradient of 72 mmHg and a right pulmonary artery *z*-score of −1.9. The CT scan showed diffuse SVAS with a critical area of 6.18 mm (Fig. 2), mild right carotid stenosis and hyperinflation of the right upper lobe of the lung.

In the operating room, after fasting for 10 h, he was premedicated with intranasal midazolam (3 mg), and monitored with pulse oximetry, blood pressure every 3 min and continuous electrocardiogram with precordial leads. Anaesthesia was induced gradually with up to 4% sevoflurane, the saphenous vein was cannulated and intravenous fentanyl (1.5 $\mu g/kg^{-1}$) and cisatracurium (0.2 mg/kg^{-1}) were administered. Three minutes later, with sevoflurane (2%), the patients presented hypotension (60/40 mmHg), ST segment depression, arterial oxygen desaturation (SpO_2: 82%) and

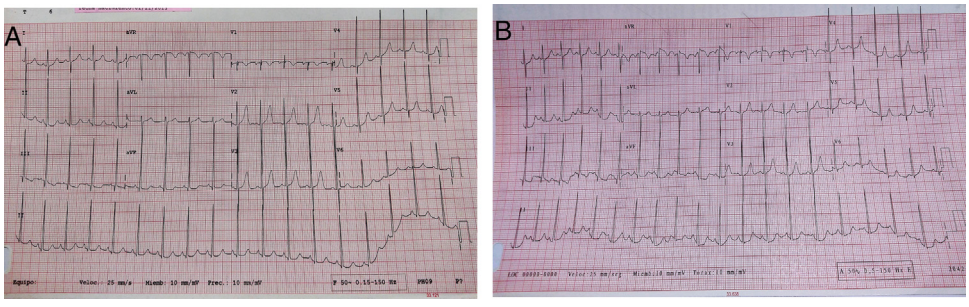

Figure 1 (A) Preoperative electrocardiogram, showing repolarisation changes with inverted T wave in lead III and signs of ventricular hypertrophy. The QT interval is within the normal range. (B) Electrocardiogram 4 months after surgery. No significant changes with respect to the previous study.
Source: With permission from the University Hospital La Paz of Madrid.