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Abstract

Uncorrected Tetralogy of Fallot (TOF) in adulthood is a rare condition and only 3% of patients reach 40 years old. Non-operated TOF patients suffer from chronic hypoxia and decreased pulmonary blood flow resulting in considerable physiological changes. The optimal management of these patients, therefore, requires a thorough understanding of the pathophysiology of the uncorrected TOF, as minor pharmacological intervention may result in severe clinical complications. We present the case of successful management of a 62-year-old man with uncorrected TOF, scheduled for a retinal detachment vitrectomy in ambulatory setting. We discuss anesthetic considerations and highlight the importance of regional anesthesia (peribulbar block), in the management of patients with severe conditions in the ambulatory setting.

Keywords: Tetralogy of Fallot, ambulatory, peribulbar block, regional anesthesia.. Authors' Address: Anesthesiology Department, Hospital de Braga, R. das Comunidades Lusíadas 133, Braga, Portugal.. Corresponding Author: : J. Barbosa, Anesthesiology Department, Hospital de Braga, R. das Comunidades Lusíadas 133, Braga, Portugal. *Email*: joao_pcb@hotmail.com

Introduction

Tetralogy of Fallot (TOF) is a cyanotic heart disease (CHD) composed by four major abnormalities: a right ventricular hypertrophy and outflow obstruction, an overriding aorta and a ventricular septal defect (1).

TOF is one of the most common causes of CHD and occurs in 3 out of 10,000 live births (2). Most patients without surgical repair would die during childhood. Survival data shows that 24% lived until the age of 10 and only 3% until 40 years old (3). Hence, a surgically unrepaired Tetralogy of Fallot in the seventh decade of life is extremely rare. There are only two documented reports of patients who survived more than 80 years without surgery (4).

Case report

A 62-year-old, 73 kg man came to preanesthetic assessment for ambulatory surgery. He presented for a left eye vitrectomy for retinal detachment. He did not take any medications and states no prior surgeries or any significant medical history. His medical records only show a prior echocardiography (dated five years before), stating an unrepaired TOF. When confronted with this information, the patient claimed that, at age of 10, he was advised to undergo surgical repair but his parents refused surgery (as himself since he became an adult) due to his asymptomatic status. He denies having chest pain or dyspnea for moderate efforts, adding that he regularly walks 10 km on weekends. Patient was classified as ASA 3 and another echocardiography was requested at this initial assessment and confirmed the unrepaired TOF showing preserved left ventricular function, mild mitral/aortic insufficiency, a large interventricular shunt with low velocity and a mean pulmonary pressure of 46mmHg. The patient was normotensive and 94% oxygen saturation on room air. Physical examination showed nail clubbing and a barrel chest. Chest auscultation showed widespread crackles. An arterial blood gas analysis, with the patient breathing spontaneously, revealed pH 7.436, pCO2 38.7 mmHg, pO2 76.5 mmHg, HCO3 25.6 mmol/L and lactic acid 0.89 mmol/L. General anesthesia was considered "high risk" and peribulbar block (PB) was chosen as the safest strategy, since in a PB, the local anesthetic is injected into the extraconal compartment, avoiding optic nerve injury and brainstem anesthesia (associated with ophthalmic regional anesthesia in the past).

Prior to surgery, the patient was monitored according to ASA standards and premedicated with 50 mcg of fentanyl and 1 mg of midazolam. A 5ml injection of 1% ropivacaine was performed at the inferotemporal quadrant of the orbital border and a second injection of 3ml at the lacrimal caruncle. A 25Gx25mm needle was used and no complications or accidents were reported with the technique. A Honan balloon was applied, and 30 mmHg pressure was kept for 15 minutes. Upon the Honan balloon removal, complete akinesia and sensitive block was obtained and surgeons were given permission to start the surgery. The surgery underwent uneventful for 90 minutes and the patient was sent to post-anesthesia care unit (PACU) and discharged home successfully 10 hours later. Follow-up telephone calls were made 24 hours, 48 hours and 1 month later and no clinical deterioration, pain or other postoperative side effects were reported.

Discussion

Regional anesthesia techniques allow a better titration, or even avoidance, of general anesthesia and a lot of cardiac and respiratory complications can be minimized in patients presenting unexpected conditions such as uncorrected TOF. This approach allowed us to safely treat the patient in an ambulatory setting, improving satisfaction at a lower cost. Submitting this patient to general anesthesia would mean a higher level of intra and postoperative monitoring with a longer hospital stay and a higher risk of hospital acquired infection and thromboembolism.

Peribulbar block provides an excellent alternative to GA in most ophthalmologic procedures and also gives prolonged satisfactory analgesia with mild systemic non-opioid drugs, minimizing the risk for post-operative nausea and vomiting as well.

Conclusion

There is scarce information about late survival in unrepaired TOF patients. We have reported the first case of ambulatory surgery in a patient with unrepaired TOF. The PB was the safest anesthetic plan for this patient with a rare and unexpected condition. The PB is a technique with low rate of complications and with a postoperative care easily managed at home. Thus, we believe that most of these patients could be done as ambulatory surgery under a PB instead of changing the surgical setting to inpatient surgery and stepping-up to higher levels of postoperative care.

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