

## Total intravenous anesthesia for an ectopia lentis repair surgery to a patient with Marfan syndrome

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### Abstract

Marfan's syndrome results some abnormalities of various organs and systems, including cardiovascular, pulmonary, musculoskeletal and ophthalmic manifestations. The anesthesiologist should be prepared for a potentially difficult intubation and unstable cardiovascular and pulmonary conditions. For our patient with Marfan syndrome scheduled for ectopia lentis repair surgery, we applied total intravenous anesthesia (TIVA) using propofol and remifentanyl for preserving the blood pressure levels stable and volume-controlled ventilation with mild peak inspiratory pressure levels for avoiding barotrauma. In this present, TIVA is recommended for children with coexisting similar abnormalities.

### Keywords

Marfan syndrome; total intravenous anesthesia; propofol; remifentanyl

### Introduction

The Marfan's Syndrome is an autosomal dominant condition caused by a mutation in the FBN1 gene on chromosome 15 that encodes the protein fibrillin. This defect results in a set of expressions of various organs and systems, being musculoskeletal, cardiovascular and ophthalmic manifestations the most notorious. It has an estimated incidence of 2 - 3 per 10,000, occurring in all racial groups [1]. Fibrillin is an important component of microfibrils, and is essential for the integrity of both elastic and non-elastic connective tissue. Abnormal fibrillin alters the elasticity and tensile strength of connective tissue, particularly in areas where fibrillin is abundant such as the proximal aorta, zonula of the lens in the eye, in long bones and the skin [2]. The diagnostic criteria used by Ghent include a family history and involvement of at least two or three organ systems if a mutation is suspected [3,4]. The cardiovascular, pulmonary, and musculoskeletal system disorders are of particular relevance to the anesthesiologist. Cardiovascular manifestations, especially aortic dilatation, aneurysm and dissection are the main cause of morbidity and mortality in MFS. Skeletal abnormalities resulting from disproportionate length of the long bones are pathognomonic of Marfan's

syndrome and this results to kyphoscoliosis. Due to excessive rib growth sternal abnormalities like pectus excavatum or carinatum may occur [5]. Pulmonary manifestations are due to pectus excavatum, pectus carinatum, kyphoscoliosis and the loss of elasticity of the lung [3-6].

To the best of our knowledge, there was no case in literature regarding total intravenous anesthesia (TIVA) for Marfan's Syndrome. Therefore, we aimed to show that TIVA is applicable as uneventfully to a patient with Marfan's Syndrome.

## Case Report

Written informed consent for publication was obtained from the patient's parent. A 14-year-old female patient with Marfan's Syndrome presented for repair of ectopia lentis surgery. She was 53 kg of weight and 177 cm of height. She had impaired vision on her right eye. Her transthoracic and transesophageal echocardiograms were reported normal. ECG showed no significant abnormalities. Performed pulmonary function test showed normal function. Her routine preoperative laboratory tests resulted normal. On examination, she was tall and thin and had long arms and fingers. She had no obvious skeletal abnormalities in terms of kyphoscoliosis and pectus carinatum or excavatum. She was positioned carefully for reducing the risk of joint and soft tissue trauma and dislocation. Monitorization with pulse oximeter, noninvasive blood pressure, electro-cardiograph and capnography was provided. Two peripheral intravenous lines were secured. Preoxygenation was performed with 100% O<sub>2</sub> for 4 min. Her anesthesia induction was carried out with propofol 2mg/kg, fentanyl 2 µg/kg and rocuronium 0.6 mg/kg. The patient was ventilated with a facemask with 100% O<sub>2</sub> for 2 min, before being intubated by direct laryngoscopy without difficulty. The choice of anesthetic maintenance type was TIVA to control blood pressure levels in desired limits. Remifentanyl infusion of 0.05-0.25 µg/kg/min rate and propofol infusion of 10 mg/kg/h for first 10 minutes, 8 mg/kg/h for next 10 minutes and 6 mg/kg/h were applied. The patient was ventilated in volume controlled ventilation. Systolic blood pressure levels were maintained between 80-110 mmHg. The surgery lasted for 45 minutes and proceeded uneventful. Reversion of neuromuscular blockade was made with sugammadex 2 mg/kg and the extubating progressed without complication. Paracetamol was administered to provide analgesia. The patient was discharged on the 2nd postoperative day without complaints.

## Discussion

It is well known that, TIVA is easily manageable and safe anesthesia method for patients coexisting with various abnormalities being cardiovascular, pulmonary, and musculoskeletal manifestations [7-9]. Therefore we chose TIVA using propofol and remifentanyl for a patient with Marfan's syndrome.

For Marfan's syndrome, the risk of perioperative morbidity and mortality, including unexplained death, is high [10]. Careful preoperative evaluation and investigation is essential [11,12]. On physical examination, one should be alert to signs of congestive heart failure. Cardiovascular functional status needs to be assessed, including ECG, cardiac catheterization, MRI and echocardiography as indicated to access the size of the aortic root and valvular function. In patients with valve replacements, antibiotic prophylaxis and

conversion from warfarin to heparin anticoagulation should be carried out timeously. In our case performed echocardiogram reported normal cardiac and vascular structures and functions.

Strict preoperative control of blood pressure is vitally important to minimize shear forces and wall stress in the aorta to decrease the risk of aortic rupture or dissection [13]. We choose to monitor blood pressure noninvasively because her transthoracic and transesophageal echocardiograms were reported normal and surgical procedure was considered as minor surgery. Perioperative maintenance of beta-blocker is recommended to reduce myocardial contractility and control aortic wall tension in susceptible patients [14,15]. Our patient wasn't on any preoperative medication and her systolic blood pressures kept between 80-110 mmHg.

Pulmonary function tests, with or without arterial blood gas should be considered if thoracic skeletal abnormalities are severe. Monitoring will vary according to functional status. Utmost care must be taken when placing arterial lines, endotracheal tubes and probes to prevent damage to weakened tissues. Regarding lung function, patients with Marfan's Syndrome normally present with restrictive ventilatory defects, not only because of the underlying emphysema, but also due to the musculoskeletal changes that affect thoracic expansion like kyphoscoliosis and pectus excavatum or carinatum [16]. The patient had no significant deformity nor pulmonary function abnormality.

The anesthesiologist should be prepared for a potentially difficult intubation owing to factors related with arched palate, retrognathia and ligamentous hyperlaxity that can lead to joint luxation during neck extension (cervical spine, temporo mandibular) [16]. Intraoperatively, careful positioning of the patient is important to prevent damage to lax joints.

Ventilatory pressures must be kept as low as possible to prevent barotrauma and reduce the risk of pneumothorax, especially if the patient has a previous diagnosis of lung cysts. Tracheomalacia has been reported as a potential complication [17]. In our case, volume-controlled ventilation with mild peak pressures was carried out and TIVA was chosen for preserving the blood pressure levels stable.

## Conclusion

In conclusion, detailed preoperative assessment of cardiovascular, pulmonary and musculoskeletal systems is essential for patients with Marfan's Syndrome that are scheduled for surgery. Appropriate and careful anesthetic technique is crucial for avoiding fatal complications. Blood pressure monitoring and control is vitally important for intraoperative management. We preferred TIVA for children with Marfan's syndrome.

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