CASE REPORT

Navigating Aortic Fragility and Seizure Risk in a Child with Marfan Syndrome Undergoing Hypospadias Repair: The Potential of Dexmedetomidine-Based Anesthesia–A Case Report

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ABSTRACT

Background: Marfan syndrome poses significant anesthetic risks, including cardiovascular instability and tissue fragility, which are further complicated in pediatric patients with epilepsy.

Case: We present the anesthetic management of a 13-year-old boy with Marfan syndrome and controlled epilepsy undergoing elective urethroplasty. To prioritize hemodynamic stability for aortic protection and prevent perioperative seizures, general anesthesia was induced and maintained with sevoflurane and a continuous dexmedetomidine infusion (loading dose 0.5 µg/kg; maintenance 0.2 µg/kg/h). This multimodal approach provided effective sympatholysis and analgesia without utilizing neuraxial blockade. Intraoperative hemodynamics remained stable, and the patient was extubated smoothly without coughing or agitation, followed by an uncomplicated discharge on the second postoperative day.

Discussion: The multimodal integration of dexmedetomidine provided effective sympatholysis, analgesia, and MAC-sparing effects. Intraoperative hemodynamics remained strictly controlled without significant bradycardia or hypotension, shielding the fragile aortic wall from elevated shear stress (dP/dt). Furthermore, dexmedetomidine mitigated the pro-convulsant risks associated with volatile anesthetics and eliminated the need for neuraxial blockade. The patient experienced a smooth, "cough-free" emergence without emergence delirium or bladder spasms. Postoperative recovery in the ward was entirely uncomplicated, with stable vital signs and no seizure activity.

Conclusion: Dexmedetomidine demonstrated efficacy as a pivotal adjunct in this complex case, enabling a non-neuraxial strategy and ensuring a smooth emergence to minimize perioperative morbidity in high-risk pediatric patients with connective tissue and neurological comorbidities

Keywords: dexmedetomidine; epilepsy; general anesthesia; hypospadias; marfan syndrome; pediatric anesthesia

INTRODUCTION

Marfan syndrome is an autosomal dominant connective tissue disorder caused by mutations in the fibrillin-1 (FBN1) gene on chromosome 15, which encodes the FBN1 protein.[1] It is among the most common inherited connective tissue disorders, with an estimated incidence of 23.2 per 100,000 population.[2] Mutations in FBN1 result in widespread connective tissue abnormalities characterized by reduced elasticity and tensile strength. The cardiovascular, ocular, and musculoskeletal systems are most frequently affected, with manifestations ranging from skeletal deformities to life-threatening cardiovascular complications.[1,3]

These systemic connective tissue abnormalities substantially increase the risk of anesthetic and surgical complications in patients with Marfan syndrome. From an anesthetic perspective, practitioners must navigate difficult airways caused by restricted mouth opening and neck mobility, manage restrictive lung physiology, and strictly control hemodynamic fluctuations to reduce aortic shear stress.[4] Tissue fragility and vascular instability pose particular challenges during surgical procedures, including the correction of congenital urogenital anomalies such as hypospadias. In addition, the coexistence of neurological comorbidities, such as epilepsy, further complicates perioperative management. The need to balance hemodynamic stability, minimize surgical stress responses, and maintain a seizure-free state necessitates careful and individualized anesthetic planning.[2,4]

Dexmedetomidine has emerged as a useful adjunct in pediatric anesthesia to address these perioperative challenges. As a highly selective α2-adrenergic agonist, dexmedetomidine provides sedation, anxiolysis, and analgesia resembling natural sleep, with minimal clinically significant respiratory depression.[5] Importantly, it attenuates sympathetic nervous system activity, contributing to stable hemodynamics and reducing perioperative tachycardia and hypertension.[6] Furthermore, dexmedetomidine has been shown to decrease anesthetic and opioid requirements and to facilitate smoother emergence from anesthesia by lowering the incidence of emergence delirium, which is particularly advantageous in pediatric patients.[6,7] These pharmacological properties suggest a potential role for dexmedetomidine in maintaining cardiovascular and neurological stability in patients with connective tissue disorders.

Anesthetic management in patients with Marfan syndrome must be highly individualized, as standardized guidelines for pediatric urological procedures in this population are currently lacking. Cardiovascular complications, including aortic root dilation and dissection, remain the primary causes of morbidity and mortality, underscoring the importance of thorough preoperative cardiovascular assessment and meticulous intraoperative hemodynamic control.[3] To the best of the authors' knowledge, reports describing the anesthetic

management of pediatric patients with Marfan syndrome and concurrent epilepsy undergoing urethroplasty are limited. Therefore, this case report presents the anesthetic management of a child with Marfan syndrome and epilepsy undergoing hypospadias repair, highlighting the perioperative considerations and the potential role of dexmedetomidine as an adjuvant anesthetic agent.

CASE

A 13-year-old boy (weight 27 kg, height 135 cm, body mass index 14.8 kg/m2) was scheduled for an elective urethroplasty for distal shaft hypospadias with chordee. His medical history was significant for Marfan syndrome and epilepsy. The diagnosis of Marfan syndrome was established clinically utilizing the revised Ghent criteria, based on a high systemic score including arachnodactyly, pectus excavatum, and thoracolumbar scoliosis.[8] He had previously undergone a chordectomy in July 2025. He had a history of epilepsy diagnosed six years prior, with the most recent seizure occurring in November 2025. The condition was well controlled with oral valproic acid (500 mg twice daily), which he had been taking consistently since September 2025. There was no history of asthma, diabetes mellitus, hypertension, or known drug or food allergies.

On physical examination, the patient was alert and cooperative, with a Glasgow Coma Scale (GCS) score of E4V5M6. Preoperative vital signs were stable: blood pressure 126/81 mmHg, heart rate 79 beats per minute, respiratory rate 20 breaths per minute, oxygen saturation 100% on room air, and body temperature 36.6°C. (Table 1) Phenotypic features consistent with Marfan syndrome were evident, including a narrow and elongated face, high-arched palate, pectus excavatum, arachnodactyly, hindfoot valgus deformity, and thoracolumbar scoliosis (Figure 1). Airway assessment revealed a Mallampati class II with adequate mouth opening and normal neck mobility (thyromental distance 3–3–2), with no predictors of difficult mask ventilation or tracheal intubation. Cardiovascular examination demonstrated a regular rhythm without murmurs or gallops, while pulmonary auscultation revealed clear vesicular breath sounds bilaterally. Preoperative laboratory investigations were within normal limits, including hemoglobin 13.7 g/dL, hematocrit 42%, leukocyte count 6,860/µL, and platelet count 440,000/µL. Coagulation parameters (PT 14.1 s, APTT 35.4 s), liver function tests (AST 23 U/L, ALT 13 U/L), renal function (urea 24.4 mg/dL, creatinine 0.72 mg/dL), and serum electrolytes (Na 137 mmol/L, K 4.5 mmol/L) were normal. Chest radiography showed no cardiomegaly but confirmed the presence of spinal scoliosis (Figure 2). Transthoracic echocardiography demonstrated normal cardiac chamber dimensions and biventricular function, with no evidence of aortic root dilatation or valvular abnormalities. The patient was classified as American Society of Anesthesiologists (ASA) physical status II. The anesthetic plan prioritized hemodynamic stability to minimize aortic wall stress associated with Marfan syndrome and the prevention of perioperative seizure activity. Premedication consisted of intravenous ondansetron (4 mg) and dexamethasone (3 mg).

Anesthesia was induced with intravenous midazolam 0.07 mg/kg (2 mg), fentanyl 1.85 µg/kg (50 µg),

propofol 1.85 mg/kg (50 mg), and dexmedetomidine 0.5 µg/kg to attenuate the sympathetic response to laryngoscopy and intubation. Neuromuscular blockade was achieved with rocuronium 0.55 mg/kg (15 mg). Tracheal intubation was performed smoothly using a 5.5-mm cuffed endotracheal tube. Anesthesia was maintained with sevoflurane 2% in a 60:40 oxygen–air mixture and a continuous infusion of dexmedetomidine at 0.2 µg/kg/h.

The surgical procedure proceeded uneventfully. Intraoperative hemodynamics remained stable throughout the operation. Fluid therapy consisted of 600 mL of Ringer's lactate. Multimodal analgesia was provided with intravenous paracetamol (400 mg) and tramadol (20 mg). Estimated blood loss was minimal (<50 mL). At the conclusion of surgery, the patient was extubated smoothly without coughing or straining and transferred to the ward. Postoperative analgesia was maintained with dexmedetomidine (0.1–0.2 µg/kg/h) and paracetamol 500 mg IV/8 hours. The postoperative course was uncomplicated, with no seizure activity or hemodynamic instability, and the patient was discharged in good condition on the second postoperative day (Table 2).

DISCUSSION

Anesthetic management in patients with Marfan syndrome necessitates a rigorous balance between profound sympatholysis to minimize aortic shear stress and the maintenance of sufficient perfusion pressure to ensure end-organ viability. The primary perioperative imperative is to dampen hemodynamic fluctuations, as the reduced tensile strength of the aortic wall in these patients makes them highly susceptible to dissection or rupture during periods of sympathetic surge.[3] This challenge was further stratified in our case by the patient's pediatric age and concurrent epilepsy, requiring a pharmacological strategy that avoided lowering the seizure threshold while ensuring hemodynamic stability. To address these multisystemic risks, we utilized dexmedetomidine as a cornerstone adjunct. Pharmacologically, dexmedetomidine is a highly selective α2-adrenergic agonist with an α2:α1selectivity ratio of 1600:1, which is eight times greater than that of clonidine.[6] This high selectivity allows for predictable sedation mediated via the locus coeruleus that parallels natural sleep, coupled with significant anxiolysis and sympatholysis, without the respiratory depression commonly associated with opioids.[5,6]

The maintenance of hemodynamic stability is critical in Marfan syndrome to minimize the rate of rise in aortic pressure (dP/dt). Current literature, which involved 48 pediatric cohorts undergoing hypospadias repair under general anesthesia, indicates that while rapid boluses of dexmedetomidine may cause transient hypertension, the predominant hemodynamic effects are a reduction in heart rate and mean arterial pressure.[6] In pediatric populations, a bradycardia of up to 30% from baseline is an expected pharmacological effect rather than a sign of toxicity.[5] In our patient, the administration of a loading dose (0.5 µg/kg) followed by a maintenance infusion (0.2 µg/kg/h)

effectively blunted the hyperdynamic response to laryngoscopy. Laryngoscopy, endotracheal intubation, and extubation provoke significant sympathetic nervous system activation characterized by transient tachycardia and hypertension.[8,9] Randomized controlled trials confirm that dexmedetomidine significantly attenuates heart rate and mean arterial pressure surges during airway instrumentation and extubation, thereby reducing the rate-pressure product and myocardial oxygen consumption.[8,9] Suppressing these acute pressor responses is critical in patients with vascular lesions or connective tissue disorders to prevent potentially hazardous arterial stress.[8]

Although bradycardia is a known side effect, recent reviews suggest that immediate intervention is rarely required unless it is accompanied by hypotension or evidence of hypoperfusion.[6] In the context of Marfan syndrome, this moderate reduction in chronotropy was therapeutically desirable to reduce myocardial oxygen demand and wall stress on the aortic root. Furthermore, regarding the patient's neurological comorbidity, dexmedetomidine offers a distinct advantage over other anesthetic agents. Unlike high-dose opioids or certain benzodiazepines, which can occasionally trigger interictal epileptiform activity, dexmedetomidine in clinical dose ranges (0.2–0. µg/kg/h) preserves neurophysiological stability and does not interfere with electrocorticography.[5] In clinical trials involving pediatric patients, intravenous dexmedetomidine has been shown to produce a dose-dependent reduction in the required minimum alveolar concentration (MAC) of sevoflurane for smooth extubation, decreasing MAC by 41% to 64% at doses of 1 to 2 µg/kg.[10] This MAC-sparing effect minimizes exposure to volatile anesthetics.[10] Furthermore, systematic reviews and meta-analyses confirm that dexmedetomidine significantly reduces seizure risk (odds ratio 0.31, $p < 0.05$) and increases the seizure threshold via alpha-2 adrenoceptor-mediated neuroprotective mechanisms without interfering with electroencephalographic monitoring.[11,12] Preclinical data also suggest that dexmedetomidine exerts neuroprotective effects via α2A-adrenoceptors, potentially attenuating neurotoxicity associated with volatile anesthetics like sevoflurane in the developing brain.[6]

Effective analgesia was paramount, as hypospadias repair is associated with significant postoperative pain and bladder spasms. While caudal epidural blockade is a standard analgesic technique for this procedure, we avoided neuraxial instrumentation due to the high prevalence of dural ectasia in Marfan syndrome. In patients with Marfan syndrome, dural ectasia causes an enlarged dural sac and increased CSF volume within the theca, which can restrict or alter the spread of intrathecally injected local anesthetics.[13,14] Clinical reports document failed spinal anesthesia requiring conversion to general anesthesia in this population.[13,14] Furthermore, spinal deformities and

meningeal diverticula increase the risk of accidental dural puncture during routine pediatric neuraxial procedures, as demonstrated by reports of accidental puncture during caudal epidural block in children with Marfan syndrome and unsuspected lumbosacral meningeal cysts.[15] Therefore, avoiding neuraxial instrumentation eliminates the risks of block failure and accidental dural puncture in patients with potential meningeal anomalies.[13–15] Instead, we relied on the systemic analgesic properties of dexmedetomidine, which are mediated through the modulation of nociceptive transmission in the dorsal horn of the spinal cord.[6] Clinical trials specifically evaluating children undergoing hypospadias repair have demonstrated that intraoperative dexmedetomidine significantly reduces the total requirements for fentanyl, morphine, and paracetamol.[6] This opioid-sparing effect was evident in our case, where the patient maintained comfort with minimal supplemental analgesia. Finally, the quality of emergence is a critical determinant of safety in patients with aortic pathology, as coughing or agitation can cause dangerous spikes in intrathoracic pressure. Sevoflurane anesthesia is associated with a high incidence of emergence delirium in children, reaching rates of up to 40%.[5] A recent meta-analysis of randomized controlled trials indicated that dexmedetomidine significantly reduces the risk of emergence delirium compared to placebo or midazolam, with a risk ratio of 0.36.[7] By maintaining the dexmedetomidine infusion until the end of surgery, we ensured a smooth, "cough-free" extubation. Postoperatively, the patient exhibited prolonged drowsiness in the ward, underscoring the known sedative tail of continuous dexmedetomidine infusion. Although this prolonged recovery serves as a favorable clinical trade-off to prevent sympathetic surges and the emergence of agitation in Marfan syndrome, it remains a notable consideration. A primary limitation of this report is the absence of a standardized objective sedation scoring to precisely measure the recovery trajectory. Additionally, as a single case report, our findings regarding the exact duration of this prolonged sedation cannot be broadly generalized, highlighting the necessity for further controlled studies to refine dosing strategies in high-risk pediatric populations.[5]

CONCLUSION

This case highlights the successful anesthetic management of a pediatric patient with Marfan syndrome and concurrent epilepsy undergoing urethroplasty. The integration of dexmedetomidine as a multimodal adjunct proved pivotal in addressing the complex dichotomy of profound sympatholysis for aortic protection and neurostability for seizure prevention. Furthermore, its analgesic efficacy facilitated a non-neuraxial strategy, avoiding potential risks associated with dural ectasia while ensuring a smooth, agitation-free emergence. This report supports the utility of a tailored, dexmedetomidine-based protocol in minimizing perioperative morbidity in high-risk syndromic children.

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Table 1. Patient hemodynamic table during surgery

Time (Waktu)	Phase (Fase)	Heart Rate (bpm)	Systolic BP (mmHg)	Diastolic BP (mmHg)	SpO2 (%)
08:00	Pre-Induction	75	124	83	100
09:01	Pre-Incision / Prep	93	100	54	100
10:08	Intraoperative	72	90	38	100
11:05	Intraoperative	68	93	44	99
11:42	Post-Extubation	73	91	49	100

Table 2. Patient hemodynamic table after surgery

Time (Waktu)	Phase (Fase)	Heart Rate (bpm)	Systolic BP (mmHg)	Diastolic BP (mmHg)	SpO2 (%)	Pain Score (NRS)
11:42	PACU Arrival	73	91	49	100	-
17:42	Ward Follow-up	65	82	43	98	0

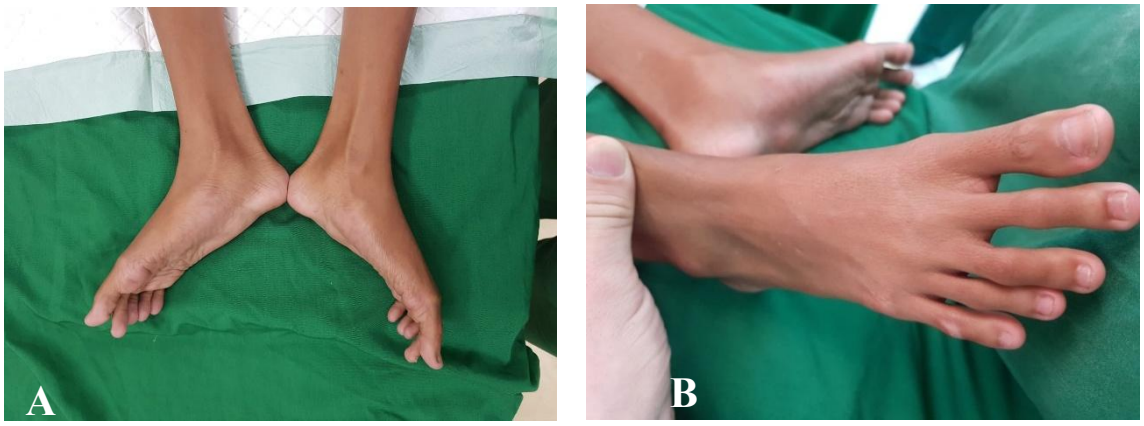


Figure 1. Clinical features characteristic of Marfan syndrome observed in the patient (A) Posterior view of the feet demonstrating bilateral hindfoot valgus deformity; (B) Lateral view revealing arachnodactyly

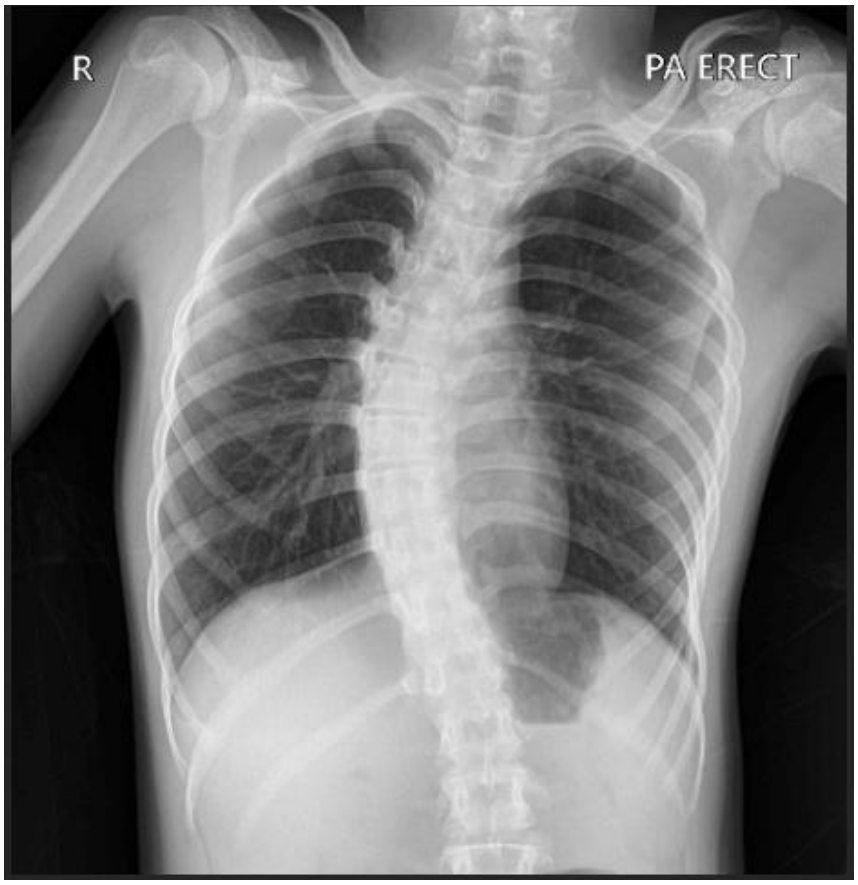


Figure 2. Preoperative chest X-ray (PA). Thoracolumbar scoliosis is noted as a musculoskeletal manifestation of Marfan syndrome.