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Anesthetic Management of a Child with Fraser Syndrome and an Atrial Septal Defect Undergoing Laparoscopic Orchidopexy: A Case Report

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A B S T R A C T

Introduction: Fraser syndrome is a rare autosomal recessive disorder with multisystem manifestations, including craniofacial, ocular, urogenital, and musculoskeletal anomalies, and it may create anesthetic challenges related to difficult airway management. The risk is magnified when congenital heart disease and laparoscopic surgery coexist, because each adds an independent physiologic burden to a child with limited reserve.

Case presentation: A 10-year-old boy weighing 8.3 kg and measuring 80 cm, with Fraser syndrome, bilateral anophthalmia, intellectual disability, a history of congenital cytomegalovirus infection, and marasmic malnutrition, underwent laparoscopic exploration of the right testis and orchidopexy of the left testis. Echocardiography demonstrated an atrial septal defect with right atrial and right ventricular dilatation; the patient was classified as American Society of Anesthesiologists physical status III. After midazolam 1 mg premedication, anesthesia was induced with fentanyl 3 mcg/kg, sevoflurane 1-2 vol%, atracurium 5 mg, and dexamethasone 4 mg, and the trachea was intubated with a 4.5-mm cuffed tube. Anesthesia was maintained with sevoflurane 1.5-2 vol% and 50% oxygen. Over the 2-hour procedure, hemodynamics were stable (blood pressure 100-130/60-68 mmHg, heart rate 82-95 beats/min) with a net positive fluid balance of 10 mL. The patient was extubated and remained stable through 24 hours.

Conclusion: Safe anesthesia in this setting rested on a layered airway plan, an understanding of shunt physiology, close hemodynamic monitoring, and measured ventilation and fluid strategies rather than on any single drug regimen. The report also highlights data elements that should be documented prospectively to strengthen comparability across similar cases.

1. Introduction

Fraser syndrome is a rare autosomal recessive multiple-malformation syndrome linked to disruption of the FRAS/FREM extracellular-matrix complex. Its clinical expression is highly variable, but ocular anomalies such as cryptophthalmos or anophthalmia, craniofacial dysmorphism, syndactyly, and renal and genital anomalies are characteristic findings. A recent newborn case report from a consanguineous family illustrates this heterogeneity well, describing bilateral anophthalmia together with syndactyly, genital anomaly, and associated cardiac abnormalities, and it underscores that the diagnosis rests on the clinical association of multiple malformations.[1] This phenotypic variability means that every patient requires individual assessment, because the particular combination of anatomical anomalies and accompanying comorbidities can differ substantially from one child to another. In children who survive into later childhood, surgical procedures for urogenital,

ophthalmologic, or other anomalies may still be required, bringing these patients into the operating room despite the rarity of the underlying condition.

Several comorbidities that cluster with syndromic disease further shape perioperative risk. A documented history of congenital cytomegalovirus infection is relevant because this is the most common congenital infection and a leading non-genetic cause of neurodevelopmental impairment; a recent multicenter cohort found that the majority of affected children carried diagnoses of neurodevelopmental conditions, including hearing loss, communication delay, and cerebral palsy. [2] When intellectual disability and severe growth restriction accompany a malformation syndrome, the anesthetic plan must account not only for anatomy but also for limited physiologic and behavioral reserve, including the reduced cooperation that complicates airway assessment and emergence.

From an anesthetic standpoint, Fraser syndrome demands particular attention to the airway. Facial disproportion, mandibular or midfacial anomalies, possible laryngeal and subglottic abnormalities, restricted mouth opening, and impaired cooperation can each complicate mask ventilation, laryngeal visualization, and tracheal intubation. Modern difficult-airway management emphasizes preoperative evaluation, the formulation of a primary plan and rescue plans, optimization of oxygenation, limitation of repeated laryngoscopy attempts, and readiness of appropriate equipment and personnel.[3] Contemporary registry data reinforce these principles. In a high-risk neonatal cohort, operator experience rather than equipment was the greatest determinant of first-attempt intubation success, and higher glottic grades reduced success.[4] Analyses of the Pediatric Difficult Intubation (PeDI) registry show that outcomes have improved over time as more structured strategies and earlier escalation have been adopted,[5] while complication rates are higher when difficult intubations are attempted at off-hours, with more than twice the odds of any complication on weekends compared with weekdays.[6] For a child with low body weight, in whom failure of oxygenation and airway trauma can progress rapidly, success on the first attempt carries special value.

Contemporary evidence also informs the tools used to achieve that first-attempt success. A systematic review and meta-analysis of randomized trials found that videolaryngoscopy improves glottic visualization across pediatric age groups and reduces esophageal intubation in the youngest children, even where first-attempt success is comparable to direct laryngoscopy in mostly normal airways,[7] and a Cochrane review in neonates reported higher first-attempt success and less airway trauma with videolaryngoscopy.[8] A pediatric randomized trial likewise showed that videolaryngoscopy produced the best glottic view and the most effective placement of a second-generation supraglottic device.[9] When direct laryngoscopy fails, registry data show that clinicians most often turn to videolaryngoscopy as a rescue technique, with success comparable to flexible bronchoscopy in most children, although flexible bronchoscopy retains an advantage in the smallest infants, underscoring the value of maintaining proficiency with both.[10] Oxygenation reserve can be extended further: a randomized trial in children demonstrated that apneic oxygenation prolongs safe apnea time, a margin that is especially valuable when laryngoscopy proves difficult.[11] The selection of a cuffed tracheal tube is supported by data showing that cuffed tubes in small children do not increase post-extubation stridor or subglottic stenosis, while providing a sealed airway suited to controlled ventilation.[12]

Perioperative risk becomes more complex in the presence of congenital heart disease (CHD). A scientific statement from the American Heart Association recommends that risk in CHD be stratified by lesion physiology, current clinical status, comorbidities, and procedural risk rather than by the diagnostic label.[13] Contemporary cohort and registry data give this principle quantitative weight. In a large multicenter registry of children with CHD undergoing noncardiac procedures, intraoperative cardiac events occurred in 5.2% of procedures, most often hypotension, with severe CHD and preoperative ventilatory support among the strongest risk factors.[14] A national surgical

database analysis spanning two eras has shown meaningful improvement in postoperative outcomes, including reduced reintubation even in severe CHD, reflecting advances in perioperative care and multidisciplinary management.[15] The pulmonary vascular bed is a particular concern: in children with pulmonary hypertension, serious adverse events are common and longer procedure duration independently increases risk, arguing for the shortest safe anesthetic.[16] Preoperative predictors of perioperative cardiac arrest or death have also been defined, supporting institution-level risk-mitigation guidelines.[17] Crucially, airway and cardiac risk are not independent: difficult tracheal intubation in patients with CHD has been associated with worse perioperative outcomes, so an effective airway plan is also a form of hemodynamic protection.[18]

Laparoscopy offers the advantages of minimally invasive surgery, but pneumoperitoneum and changes in patient position can affect venous return, intrathoracic pressure, ventilation, and carbon dioxide homeostasis. In a child with CHD, these changes must be anticipated through coordination between the anesthesiologist and the surgeon, titration of anesthesia, controlled ventilation, monitoring proportional to risk, and a careful fluid strategy. National cohort data indicate that children with CHD do undergo laparoscopic surgery requiring abdominal insufflation and that selected patients can do well, with outcomes influenced by lesion complexity.[19] Superimposed marasmic malnutrition adds a further modifier, as wasting has been independently associated with higher odds of postoperative complications in pediatric abdominal surgery.[20]

The novelty of this report lies in the simultaneous convergence of risks that are rarely described together in a single child: Fraser syndrome with severe ocular anomalies, intellectual disability, marasmic malnutrition, a urogenital anomaly requiring laparoscopy, and acyanotic CHD with right-heart dilatation. The aim of this report is to describe the anesthetic management of this combination, to analyze the decisions made against current evidence, and to make explicit the data elements whose documentation would strengthen the lessons that such cases can offer.

2. Case Presentation

Written informed consent for publication of this case report and the accompanying clinical image was obtained from the patient's parents. A 10-year-old boy with a diagnosis of Fraser syndrome was scheduled for laparoscopic exploration of the right testis and orchidopexy of the left testis. His body weight was 8.3 kg and his height was 80 cm, indicating severe growth restriction. Recorded comorbidities included bilateral anophthalmia, intellectual disability, a history of congenital cytomegalovirus infection, and marasmic malnutrition. The surgical indication was a non-palpable testis; laparoscopic exploration was planned to assess the right testis, and orchidopexy was performed on the left testis. These baseline characteristics and the full perioperative course are summarized in Table 1.

Cardiovascular examination revealed a pansystolic murmur at the second intercostal space. Preoperative echocardiography demonstrated an ASD with right atrial and right ventricular dilatation. It is worth noting that a pansystolic murmur is not the classic auscultatory finding of an isolated secundum ASD, which typically produces an ejection systolic murmur with fixed splitting of the second heart sound; this discrepancy is one reason the precise lesion subtype could not be confirmed from the available record. Detailed data on pulmonary arterial pressure, the pulmonary-to-systemic flow ratio, right ventricular function, and the degree of valvular regurgitation were not available in the records reviewed for this report. As shown in Figure 1, the preoperative anteroposterior chest radiograph demonstrated an enlarged cardiac silhouette, predominantly toward the left (Figure 1, label 1), together with increased bilateral perihilar bronchovascular markings (Figure 1, label 2); these appearances are consistent with increased pulmonary blood flow and reinforced the need for vigilance regarding right-heart volume loading throughout the perioperative period.

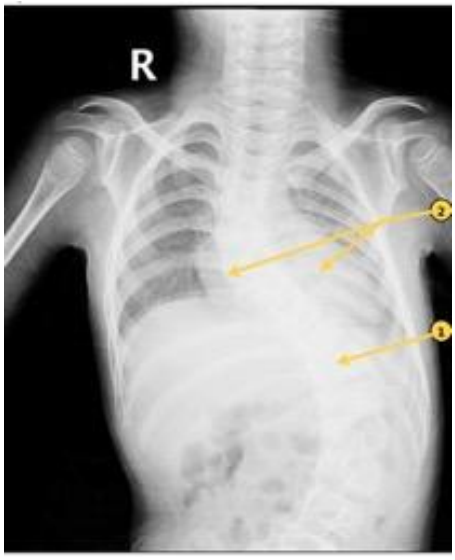

Figure 1. Preoperative frontal (anteroposterior) chest radiograph. (1) The cardiac silhouette is enlarged, predominantly toward the left; (2) bilateral perihilar bronchovascular markings are increased. These appearances are consistent with increased pulmonary blood flow in a child with congenital heart disease and should be correlated with echocardiography and the formal radiology report. The image is de-identified; patient identity is not shown.

The patient was taking piracetam, vitamin B6, folic acid, spironolactone, bisoprolol, and furosemide on a chronic basis, as listed in Table 1. On the basis of his systemic condition, his physical status was classified as American Society of Anesthesiologists (ASA) physical status III. The anesthetic plan was general anesthesia with controlled ventilation through an endotracheal tube. The history of Fraser syndrome and craniofacial anomalies was treated as a risk factor for a difficult airway, although details of the preoperative airway assessment, such as mouth opening, Mallampati classification, thyromental distance, and laryngoscopy findings, were not recorded in the reviewed documentation. Accordingly, appropriately sized facemasks, supraglottic airway devices, a videolaryngoscope, tracheal tubes of several sizes, and a clear assignment of team roles were prepared as part of risk mitigation before induction.

Before transfer to the operating room, the patient received midazolam 1 mg. Anesthesia was induced with fentanyl 3 mcg/kg and titration of sevoflurane 1-2 vol%. Atracurium 5 mg was given to provide muscle relaxation, and dexamethasone 4 mg was administered according to the perioperative record. The trachea was intubated with a cuffed endotracheal tube of 4.5 mm internal diameter, and tube position was confirmed by bilateral auscultation, as detailed in Table 1. The number of intubation attempts, the laryngoscopy device used, the laryngeal view grade, and any use of alternative airway adjuncts were not documented in the available material; this absence is addressed in the Discussion as a reporting limitation rather than as evidence of an easy airway.

A nasogastric tube was placed for gastric decompression, a measure that also helps limit diaphragmatic splinting during pneumoperitoneum. Anesthesia was maintained with sevoflurane 1.5-2 vol% and 50% oxygen. During the procedure, which lasted approximately 2 hours, systolic blood pressure ranged from 100 to 130 mmHg, diastolic blood pressure from 60 to 68 mmHg, and heart rate from 82 to 95 beats per minute; the net fluid balance was positive at 10 mL, as detailed in Table 1. No hemodynamic instability was reported. Continuous pulse oximetry, capnography, and electrocardiography were used as standard intraoperative monitors; however, the recorded ranges of peripheral oxygen saturation, end-tidal carbon dioxide, peak airway pressure, and insufflation pressure were not available in the documentation reviewed for this report, a limitation addressed in the Discussion. At the end of the procedure, the patient was extubated after meeting

standard recovery criteria, with airway equipment available for reintubation, and was transferred to the recovery room with stable vital signs. Evaluation at 24 hours after anesthesia showed that the clinical and hemodynamic condition remained stable. The perioperative course can be summarized as a simple timeline: premedication before transfer to the operating room, induction and tracheal intubation, an approximately 2-hour laparoscopic procedure under maintained hemodynamic stability, extubation in the operating room, and confirmed clinical and hemodynamic stability at 24 hours.

Table 1. Summary of clinical data and anesthetic management.

Domain	Details
Patient characteristics	Male, 10 years; body weight 8.3 kg; height 80 cm; severe growth restriction.
Diagnosis and comorbidities	Fraser syndrome; bilateral anophthalmia; intellectual disability; history of congenital cytomegalovirus infection; marasmic malnutrition; atrial septal defect with right atrial and right ventricular dilatation.
Surgical procedure	Laparoscopic exploration of the non-palpable right testis and orchidopexy of the left testis.
Preoperative findings	Pansystolic murmur at the second intercostal space; ASA physical status III; chronic medications: piracetam, vitamin B6, folic acid, spironolactone, bisoprolol, furosemide.
Premedication	Midazolam 1 mg before transfer to the operating room.
Induction	Fentanyl 3 mcg/kg, sevoflurane 1-2 vol%, atracurium 5 mg, dexamethasone 4 mg.
Airway management	Cuffed endotracheal tube, 4.5 mm internal diameter; position confirmed by bilateral auscultation; nasogastric tube placed for gastric decompression.
Maintenance	Sevoflurane 1.5-2 vol% with 50% oxygen; controlled ventilation.
Intraoperative course	Duration approximately 2 hours; blood pressure 100-130/60-68 mmHg; heart rate 82-95 beats/min; net positive fluid balance 10 mL; no hemodynamic instability reported.
Early outcome	Successful extubation; stable vital signs in recovery and through 24 hours after anesthesia.

Notes: ASA: American Society of Anesthesiologists.

3. Discussion

This report describes general anesthesia in a child with a combination of risks that is rarely reported together: Fraser syndrome, severe ocular anomalies, intellectual disability, marasmic malnutrition, a urogenital anomaly, and acyanotic CHD with right-heart dilatation. The instructive value of the case lies not simply in successful intubation or in intraoperative blood-pressure stability, but in the need to integrate assessment of airway anatomy, shunt physiology, the effects of pneumoperitoneum, and the limited physiologic reserve imposed by severe growth restriction. The report should therefore be read as an example of individualized, risk-based decision-making rather than as proof that any single anesthetic regimen can be generalized to all patients with Fraser syndrome.

Phenotype-driven, not label-driven, airway assessment

The evidence base for Fraser syndrome is dominated by case reports, so the precise frequency of difficult airways, the spectrum of laryngotracheal anomalies, and anesthetic outcomes cannot be estimated uniformly. The recent newborn case report cited above confirms that the phenotype can combine anophthalmia with craniofacial, skeletal, genital, and cardiac anomalies, and that presentation is highly heterogeneous.[1] The practical consequence is that anesthetic risk should not be predicted from the syndrome label alone. Assessment must translate the patient's actual phenotype, including facial anatomy, mandibular and cervical mobility, mouth opening, a history of stridor or recurrent airway infection, prior anesthetic history, and any suspicion of laryngeal or subglottic anomaly, into a specific airway plan. In this

child, the additional burden of intellectual disability and a history of congenital cytomegalovirus infection, both associated with neurodevelopmental impairment, further limited cooperation and reinforced the need for a controlled, well-prepared induction.[2]

In this patient, bilateral anophthalmia and a history of a multiple-malformation syndrome were sufficient reasons to regard the airway as potentially difficult, even though Mallampati score, thyromental distance, mouth opening, and laryngoscopy findings were not available. The ASA practice guidelines emphasize the importance of identifying risk, optimizing oxygenation, formulating initial ventilation and intubation plans, preparing rescue plans, and planning extubation before anesthesia begins.[3] In a child with a syndromic disorder, readiness of correctly sized masks, supraglottic devices, a videolaryngoscope, a flexible bronchoscope where available, tracheal tubes in several sizes, and clearly assigned team roles is part of risk mitigation; it does not imply that every device must be used routinely. The literature on rescue techniques supports keeping more than one option ready, since videolaryngoscopy and flexible bronchoscopy each retain specific advantages after failed direct laryngoscopy.[10]

Lessons from the pediatric difficult-airway registries and device evidence

Registry data strengthen the rationale for limiting intubation attempts and avoiding persistence with an ineffective technique. A high-risk neonatal cohort found that operator experience, rather than equipment, was the principal determinant of first-attempt success, and that higher glottic grades reduced it.[4] Analyses of the PeDI registry show outcomes improving as structured strategies and earlier escalation are adopted,[5] and that complications are more frequent when difficult intubations occur at off-hours, with more than double the odds of any complication on weekends.[6] The contemporary device literature complements these findings: a meta-analysis of randomized trials shows that videolaryngoscopy improves glottic visualization and reduces esophageal intubation in infants,[7] a Cochrane review reports higher first-attempt success and less trauma in neonates,[8] and a pediatric randomized trial confirms superior glottic views and device placement with videolaryngoscopy.[9] In a child with limited oxygen reserve, the demonstrated ability of apneic oxygenation to prolong safe apnea time provides an additional safety margin during a potentially prolonged laryngoscopy.[11] In the present case, intubation with a 4.5-mm cuffed tube was successful and was not followed by any reported complication; the choice of a cuffed tube is consistent with evidence that cuffed tubes do not increase post-extubation stridor in small children while supporting a sealed airway for controlled ventilation.[12] Nevertheless, success alone does not characterize the degree of airway difficulty. Because the number of attempts and the laryngeal view were not documented, the airway in this case cannot be retrospectively classified as either easy or difficult; this is the single most important missing airway datum, and naming it is more honest than inferring an easy airway from an uncomplicated result.

The choice of an inhalational induction with sevoflurane in an anticipated difficult airway deserves explicit framing as a deliberate trade-off. An inhalational induction can preserve the option of spontaneous ventilation and is often pragmatic when intravenous access or patient cooperation is limited, as may occur in a child with intellectual disability; the counterbalancing risk is that deepening volatile anesthesia can provoke airway obstruction or laryngospasm before the airway is secured. The conventional cautious sequence in this situation is to confirm the ability to ventilate before administering a non-depolarizing neuromuscular blocker, and to have rescue equipment and personnel immediately available, as was prepared in this case.[3] Making this reasoning explicit is intended to convey the decision logic to readers rather than to present the drug list as a protocol.

Stratifying congenital heart disease by physiology

The CHD in this case adds a dimension of risk distinct from the airway problem. In an ASD, the magnitude and direction of the shunt are influenced

by the size of the defect, ventricular compliance, pulmonary and systemic vascular resistance, and the presence of pulmonary hypertension or ventricular dysfunction. Right atrial and right ventricular dilatation on echocardiography point to right-heart volume loading, but without data on pulmonary arterial pressure, the pulmonary-to-systemic flow ratio, and ventricular function, the degree of shunt physiology cannot be classified quantitatively. The American Heart Association scientific statement recommends that stratification be based on lesion physiology, current clinical status, comorbidities, and procedural risk, rather than on the cardiac diagnostic label alone.[13] Contemporary data make this concrete: in a multicenter registry, intraoperative cardiac events occurred in 5.2% of noncardiac procedures in children with CHD, most often hypotension, and the risk rose sharply with severe CHD and with preoperative ventilatory support.[14] Reassuringly, a national database analysis across two eras has documented improving outcomes, including reduced reintubation even in severe CHD, which reflects the value of risk stratification and multidisciplinary care.[15]

Several lines of evidence support tailoring management to this patient's physiology, as summarized in Table 2. Preoperative predictors of perioperative cardiac arrest or death have been identified and can be incorporated into institutional guidelines,[17] and studies of children with pulmonary hypertension show that serious adverse events are common and that longer procedure duration independently increases risk, which argues for minimizing both anesthetic and pneumoperitoneum time.[16] For this patient, an age of 10 years did not automatically imply low risk, because very low body weight, nutritional impairment, a potentially difficult airway, and an ASD with right-heart dilatation all reduce tolerance for physiologic perturbation.

Table 2. Synthesis of recent evidence mapped to decision-making in this case.

Clinical domain	Key evidence	Implication for this case
Fraser syndrome spectrum	A recent newborn case report confirms wide phenotypic heterogeneity, consanguinity, and the co-occurrence of anophthalmia with cardiac and genital anomalies.	Anesthetic planning must be based on the individual phenotype, not on the syndrome label alone.
Pediatric difficult airway	Registry cohorts show operator experience outweighs equipment for first-attempt success, complications rise with off-hours timing, and videolaryngoscopy improves the glottic view.	Alternative devices and a rescue plan must be ready before induction, with first-attempt success prioritized by an experienced operator.
CHD for noncardiac surgery	A multicenter registry reports intraoperative cardiac events in 5.2% of procedures, with severe CHD and preoperative ventilation as strong risk factors; outcomes have improved across eras.	An atrial septal defect with right-heart dilatation requires control of preload, afterload, and pulmonary vascular resistance, and vigilance for hypotension.
Pulmonary vascular burden	In children with pulmonary hypertension, serious adverse events are common and longer procedure duration independently increases risk.	Minimize anesthesia and pneumoperitoneum time and avoid factors that raise pulmonary vascular resistance.
Airway-CHD interaction	Difficult tracheal intubation in patients with CHD is associated with worse perioperative outcomes.	Airway optimization is itself a hemodynamic-protective measure, not a separate problem.
Laparoscopy and host modifiers	Cohort data support laparoscopy in selected children with CHD, while wasting independently raises the odds of postoperative complications.	Use the lowest effective insufflation pressure and physiology-targeted ventilation; apply weight-based dosing and thermal vigilance.

Notes: CHD: congenital heart disease.

Practically, the goal of anesthesia in a patient with right-heart volume loading is to avoid abrupt changes in preload, afterload, contractility, and pulmonary vascular resistance. Hypoxia, hypercarbia, acidosis, hypothermia, uncontrolled pain, and high airway pressures can all raise pulmonary vascular resistance and burden the right ventricle, whereas a sharp fall in preload or a large change in systemic vascular resistance can compromise effective forward flow. [13,16] The chronic use of spironolactone, furosemide, and bisoprolol in this patient required individualized assessment of volume status, heart rate, blood pressure, and routine therapy; these medications cannot, by themselves, be taken as proof of active heart failure without additional clinical data. The stable heart rate of 82 to 95 beats per minute and the maintained blood pressure of 100-130/60-68 mmHg recorded during the procedure, as detailed in Table 1, are consistent with an anesthetic conducted to preserve these targets, although they do not substitute for direct measures of cardiac output or perfusion.

Shunt physiology of the atrial septal defect and its anesthetic translation

An ASD produces a communication at the atrial level through which blood shunts according to the relative compliance of the two ventricles and the relative resistances of the pulmonary and systemic circulations. In the typical secundum defect, the more compliant right ventricle and the lower pulmonary vascular resistance favor a left-to-right shunt, which over time imposes a chronic volume load on the right atrium and right ventricle and increases pulmonary blood flow. The right atrial and right ventricular dilatation demonstrated on this patient's echocardiography, and the radiographic signs of increased pulmonary blood flow shown in Figure 1, are the structural and imaging signatures of that chronic volume load. The magnitude of the shunt is commonly expressed as the pulmonary-to-systemic flow ratio; a clinically significant left-to-right shunt is associated with progressive right-heart enlargement and, in some patients over years, with elevation of pulmonary vascular resistance. Because the quantitative shunt data were not available for this child, the anesthetic plan had to assume a physiologically meaningful defect and protect the right heart accordingly.[13,16]

Translating this physiology to the operating room means that the anesthesiologist manipulates the balance between systemic and pulmonary vascular resistance with every intervention. Maneuvers or drugs that sharply raise pulmonary vascular resistance, including hypoxia, hypercarbia, acidosis, hypothermia, pain, and excessive airway pressure, increase the afterload on an already volume-loaded right ventricle and can, in defects with elevated pulmonary pressures, promote reversal of the shunt toward right-to-left flow with consequent hypoxemia. Conversely, a precipitous fall in systemic vascular resistance can increase left-to-right shunting and reduce effective systemic output, while abrupt reductions in preload can underfill a right ventricle that depends on adequate filling to maintain stroke volume.[13,14] The practical targets that follow are maintenance of normoxia and normocapnia, avoidance of acidosis and hypothermia, preservation of heart rate and contractility, and the avoidance of large, sudden swings in either vascular bed. The hemodynamic stability recorded in this case is consistent with an anesthetic conducted within these constraints, even though it cannot confirm the adequacy of cardiac output by itself.

Paradoxical embolism and meticulous air precautions

Any atrial-level communication permits paradoxical embolism, in which air or particulate matter from the venous side crosses to the systemic circulation and can reach the coronary or cerebral beds. Even a predominantly left-to-right shunt can transiently reverse during coughing, straining, Valsalva, or the swings in intrathoracic pressure that accompany positive-pressure ventilation and pneumoperitoneum, so the presence of a left-to-right ASD does not eliminate the risk.[13] The defensive measures are simple but must be deliberate: careful de-airing of all intravenous lines, the use of air filters where available, avoidance of air bubbles in syringes and infusions, and prompt attention to any sudden, unexplained desaturation or hemodynamic change

during insufflation. Laparoscopy adds a specific concern because carbon dioxide used for insufflation can, in rare circumstances, enter the venous system; vigilance for the abrupt fall in end-tidal carbon dioxide and hemodynamic collapse that signal gas embolism is therefore part of safe conduct, and these monitored parameters should be documented.[19]

Airway and cardiac risk are a single problem

Studies linking CHD and the airway are directly relevant to this case. A retrospective study in patients with CHD found that difficult tracheal intubation was associated with less favorable perioperative outcomes, so airway risk and cardiac risk should not be treated as two separate problems.[18] Conceptually, episodes of hypoxemia, hypercarbia, or sympathetic stimulation provoked by airway manipulation can affect cardiac physiology and narrow the safety margin in a patient with limited reserve, and the same perturbations raise pulmonary vascular resistance and burden the right ventricle.[16] An effective airway plan is therefore part of hemodynamic protection, and the preparation for alternative devices described above served both the airway and the circulation in this child.

Pharmacologic rationale of the chosen agents

The agents used in this case can each be justified on physiologic grounds while acknowledging that doses must be referenced to the child's actual low body weight of 8.3 kg. Midazolam 1 mg as premedication provides anxiolysis and smooths separation and transfer, which is valuable in a child with intellectual disability in whom cooperation is limited; the trade-off is the potential for sedation and airway compromise before the airway is secured, so a small titrated dose with monitoring is appropriate in a patient with a potentially difficult airway. [3] Fentanyl 3 mcg/kg blunts the sympathetic response to laryngoscopy and surgical stimulation and supports hemodynamic stability, an advantage in a heart that tolerates tachycardia and hypertensive surges poorly, while its lack of significant direct myocardial depression makes it well suited to CHD. [13]

Sevoflurane offers a smooth inhalational induction that is particularly useful when intravenous access is difficult or when a child cannot cooperate with an intravenous induction, and it allows the depth of anesthesia to be titrated rapidly. Its dose-dependent reduction in systemic vascular resistance and its mild myocardial depression are the principal cautions in a patient with a shunt, because a marked fall in systemic vascular resistance can alter shunt balance; careful titration to the lowest effective concentration, reflected in the 1.5-2 vol% maintenance range used here, mitigates this concern, and studies in children with pulmonary hypertension found that the choice of anesthetic technique itself was not associated with adverse events when conducted carefully. [16] Atracurium is an attractive neuromuscular blocking choice in this context because its Hofmann elimination is independent of hepatic and renal function, an advantage in a malnourished child in whom organ reserve and drug handling may be unpredictable; the recognized caution is histamine release with rapid or large bolus dosing, which is avoided by measured administration. Dexamethasone 4 mg contributes prophylaxis against postoperative nausea and vomiting and may reduce airway edema after instrumentation, a relevant consideration when the airway is potentially difficult and reintubation would be hazardous; a recent randomized trial of laparoscopic surgery reaffirmed dexamethasone as part of effective multimodal antiemetic prophylaxis, and notably identified added benefit from pyridoxine, a drug this patient already received chronically. [21] Across all of these agents, dosing to actual body weight, with attention to clinical response rather than to age-based assumptions, is the unifying safety principle. [20]

Physiologic consequences of laparoscopy

Laparoscopy provides minimally invasive access, but pneumoperitoneum can reduce venous return by increasing intra-abdominal pressure, raise intrathoracic pressure during controlled ventilation, and increase carbon dioxide absorption. These changes can modify cardiac output and pulmonary vascular resistance, particularly in a child with CHD. National cohort data confirm that children with CHD, including those with single-ventricle physiology, do undergo laparoscopic procedures requiring

insufflation, that selected patients have acceptable outcomes, and that length of stay may be shorter than with open surgery, while outcomes still track with cardiac complexity. [19] In this case, the implications were a need for the lowest insufflation pressure that still provided an adequate operative field, limitation of pneumoperitoneum duration where possible, ventilation aimed at maintaining normocapnia, and active communication between the surgeon and the anesthesiologist when hemodynamic or ventilatory changes occurred. The need to limit duration is reinforced by evidence that, in children with pulmonary vascular vulnerability, longer procedures independently increase the risk of serious adverse events. [16] The insufflation pressure, patient position, end-tidal carbon dioxide, serial oxygen saturation, peak airway pressure, tidal volume, and respiratory rate were not recorded in the available data, and these parameters should be regarded as essential elements for future reporting rather than values that can be reconstructed after the procedure.

Malnutrition, thermoregulation, and fluid management

Marasmic malnutrition and growth failure are additional risk modifiers that cannot be separated from the interpretation of drug dosing and monitoring. A body weight of 8.3 kg at 10 years of age requires that device sizes and drug doses be chosen according to actual body weight and clinical response, while anticipating limited energy reserve and impaired thermoregulation. Children are especially prone to intraoperative hypothermia because of their high surface-area-to-weight ratio, immature thermoregulatory centers, and thin subcutaneous fat, and structured perioperative insulation is recommended to prevent it; in a malnourished child these vulnerabilities are amplified, making active warming with forced-air devices and warmed fluids a cardiac as well as a thermal intervention, since hypothermia raises pulmonary vascular resistance.[16,22] The pediatric surgical literature has shown that wasting is independently associated with higher odds of postoperative complications, which means that poor nutritional status should heighten vigilance even when surgery is elective.[20] Depleted glycogen stores and chronic diuretic therapy further argue for individualized assessment of glucose, electrolytes, and volume status, and documentation of hemoglobin, glucose, electrolytes, renal function, temperature, fluid type, and urine output would enrich the assessment of anesthetic safety.

A net positive fluid balance of 10 mL and stable hemodynamics over 2 hours, as detailed in Table 1, indicate that a measured fluid approach was achievable in this case. However, a single fluid-balance figure does not capture the whole perfusion picture; fluid type, input and output volumes, blood loss, urine output, and clinical assessment of perfusion remain necessary. This is consistent with a perioperative approach to CHD that emphasizes monitoring and therapy based on the individual patient's physiology rather than the use of uniform volume targets. [13,14]

Postoperative analgesia and the case for opioid-sparing strategies

Although postoperative analgesic details were not documented in the available record, the physiology of this patient argues strongly for a multimodal, opioid-sparing plan. Effective analgesia is itself hemodynamic protection, because uncontrolled pain raises sympathetic tone and pulmonary vascular resistance and can destabilize a volume-loaded right heart; at the same time, systemic opioids carry a risk of respiratory depression and hypercarbia that is especially hazardous in a child with a potentially difficult airway and a shunt lesion.[16,18] For laparoscopic urologic surgery, regional techniques such as a caudal block or other fascial-plane approaches, combined with paracetamol and careful local infiltration by the surgeon, can provide analgesia while minimizing opioid exposure, and the multimodal antiemetic strategy validated in recent laparoscopic-surgery trials complements this approach.[21-23] The choice must be individualized against coagulation status, anatomy, and the risks of each technique, but the principle of limiting respiratory-depressant load while controlling the sympathetic response is well aligned with the goals of CHD anesthesia.

Emergence, recovery, and a proposed minimum dataset

The extubation phase should be understood as part of the difficult-airway strategy. The ASA guidelines incorporate oxygenation, assessment of the risk of obstruction or edema, readiness for reintubation, and planning of the recovery location into this phase. [3] In this case, the patient was extubated after meeting standard recovery criteria, with reintubation equipment available, and remained stable through 24 hours after anesthesia, which is a favorable early outcome. However, the oxygen requirement, the method of postoperative monitoring, pain scores, the occurrence of nausea and vomiting, signs of airway obstruction, and the criteria for discharge from recovery were not available. A recurring theme of this discussion is that the instructive value of a complex case is limited less by what was done than by what was recorded. To make reports of this kind comparable and actionable, a minimum dataset should be captured prospectively: for the airway, the Mallampati class, mouth opening, thyromental distance, the number of intubation attempts, the initial and rescue devices, the laryngeal view, and oxygenation during intubation; for the heart, the confirmed defect subtype, pulmonary arterial pressure, the pulmonary-to-systemic flow ratio, and ventricular function; for ventilation and laparoscopy, the insufflation pressure and duration, patient position, end-tidal carbon dioxide, peak airway pressure, tidal volume, and respiratory rate; for metabolism, hemoglobin, glucose, electrolytes, renal function, and temperature; and for recovery, oxygen requirement, pain scores, nausea and vomiting, and discharge criteria.[3,14,20]

Taken together, the synthesis of the literature, as mapped in Table 2, shows a convergence of messages from recent work on Fraser syndrome, the pediatric airway registries, noncardiac CHD studies, and laparoscopy research: success depends on early risk identification, strategies tailored to physiology, limitation of exposure to predictable perturbations, and readiness to escalate.[1,4,14,19,20] This case supports that approach, but the absence of detailed data, particularly a confirmed ASD subtype, pulmonary pressures, ventricular function, airway evaluation, ventilation and pneumoperitoneum parameters, and longer-term postoperative outcomes, limits generalization. The conclusions of this report should therefore remain bounded to the short-term success of management in a single patient.

4. Conclusion

Anesthetic management of a child with Fraser syndrome and acyanotic CHD undergoing laparoscopy requires an individualized, multidisciplinary approach. Airway risk arising from the syndromic anomalies must be anticipated through preoperative assessment, a primary plan together with rescue plans, and readiness of difficult-airway equipment. The finding of an ASD with right-heart dilatation demands maintenance of stable hemodynamics, ventilation that avoids factors which increase pulmonary vascular resistance, and measured fluid management. It is important to separate what can and cannot be generalized from this report: the management principles described, namely physiology-based risk stratification, a layered airway plan, normocapnic controlled ventilation, the lowest effective insufflation pressure, weight-based dosing, and thermal and metabolic vigilance, are transferable to similar patients, whereas the favorable outcome itself is that of a single child and cannot be generalized. In this case, general anesthesia with tracheal intubation and controlled ventilation proceeded without hemodynamic instability, followed by extubation and a good early outcome through 24 hours. More complete documentation of airway, echocardiographic, ventilation, pneumoperitoneum, and postoperative monitoring data, ideally captured prospectively as the minimum dataset proposed here, remains necessary so that the instructive value of such case reports can be strengthened.

Declarations

Ethics approval and consent to participate

Written informed consent for publication of this case report and the accompanying clinical image was obtained from the patient's parents. The report was

prepared in accordance with institutional ethical standards for case reports and the principles of the Declaration of Helsinki.

Consent for publication

Written informed consent for publication of the clinical details and the de-identified image was obtained from the patient's parents.

Availability of data and materials

The clinical data supporting this report are contained within the article. Further details are available from the corresponding author on reasonable request, subject to protection of patient confidentiality.

Competing interests

All authors declare that they have no competing interests.

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Authors' contributions

All authors contributed to the clinical management, drafting, and critical revision of the manuscript, and all authors have reviewed and approved the final version.

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