

Perioperative Risk Assessment and Anaesthetic Considerations in Cleft Lip Repair for an Infant with Dextrocardia: Strategies in the Absence of Genetic Workup

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ABSTRACT

Background: Cleft lip repair is usually safe in healthy infants, but the presence of dextrocardia raises perioperative concerns because of its frequent association with congenital heart disease and syndromes. In resource-limited settings, clinicians must make decisions without genetic testing or advanced imaging.

Case Presentation: A 3-month-old male infant weighing 5.3 kg presented with a complete right unilateral cleft lip. A chest radiograph revealed dextrocardia. Echocardiography confirmed dextrocardia with situs solitus, normal atrioventricular and ventriculoarterial concordance, preserved biventricular function, and no structural defects (no ASD, VSD, or PDA). Abdominal ultrasound confirmed situs solitus. Nutritional assessment showed mild undernutrition related to feeding difficulty. After nutritional optimization and multidisciplinary evaluation, the surgical team proceeded with primary cleft lip repair under general anesthesia. The procedure used the Fisher technique and was completed without intraoperative complications. The patient recovered uneventfully, resumed oral feeding within 12 hours, and showed good wound healing and weight gain on follow-up.

Conclusion: Isolated dextrocardia does not preclude safe cleft lip repair when echocardiography rules out structural heart disease. In settings without genetic testing, clinicians can achieve good outcomes through structured clinical evaluation, basic imaging, and close multidisciplinary coordination. This case demonstrates how careful perioperative planning and tailored anaesthetic strategies ensure safe surgical repair in a resource-limited environment.

KEYWORDS: Cleft lip, dextrocardia, perioperative risk, paediatric anaesthesia, resource-limited setting.

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INTRODUCTION

Cleft lip and dextrocardia are congenital anomalies that often co-occur in various genetic syndromes are associated with child developmental challenges. Cleft lip is among the most common congenital craniofacial anomalies, with an estimated global prevalence of 1 in 700 live births. Early surgical repair is essential to improve feeding, speech development, and psychosocial integration. Surgical timing is usually guided by the "Rule of 10s" (10 weeks of age, 10 pounds in weight, and hemoglobin of 10 g/dL), provided the infant is otherwise medically fit (1,2). Cleft lip, typically visible at birth, results

from a failure of fusion of the lips during embryonic development, resulting in a gap or cleft. Dextrocardia, defined as a right-sided location of the cardiac apex, occurs in approximately 1 in 12,000 live births. While it may present as an isolated anomaly, dextrocardia is more often associated with congenital heart disease or syndromic conditions such as heterotaxy or trisomy 13 (3). Furthermore, this creates additional complexity for anaesthetic management during pediatrics surgery, as intracardiac shunts or abnormal great vessel anatomy may increase perioperative risk.

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Based on anatomical considerations, the anaesthetic management of patients with cleft lip and dextrocardia is particularly complex. Airway challenges in cleft lip/palate patients are primarily due to orofacial anatomy (e.g., inability to obtain a mask seal, palatal clefting), micrognathia, or coexisting syndromic features. Therefore, routine airway assessment and plan should focus on cleft-related airway issues rather than cardiac position alone. Cardiac malposition does, however, necessitate adaptation of monitoring (e.g., ECG lead placement) and awareness of possible associated cardiac anomalies (4). A systematic approach to perioperative risk assessment, including evaluation of cardiac function and respiratory status, is therefore essential to mitigate these potential complications (5).

A variety of genetic syndromes involving cleft lip and dextrocardia have been identified. For example, in Meckel-Gruber Syndrome (MKS), anomalies including cleft lip and/or palate, laterality defects, and congenital cardiovascular malformations such as dextrocardia are frequently observed (6). The presence of cleft lip alongside dextrocardia has been documented in multiple cases where congenital anomalies co-occur with genetic abnormalities, underscoring their link to syndromic conditions rather than isolated occurrences (7). In trisomy-associated conditions, cleft lip is one of the most prevalent craniofacial malformations. It occurs in approximately 45% of cases with trisomy 13, where cardiac defects including dextrocardia also present at a significant frequency (8,9). This research aims to find the practical strategies for perioperative evaluation and anaesthetic planning to resource-limited settings, specifically assessing the unique challenges associated with managing cleft lip repairs and dextrocardia to optimize surgical outcomes and patient safety.

Cleft lip is one of the most prevalent congenital malformations, often requiring surgical intervention during infancy. The timing of this surgery is crucial, as both early and late repairs come with their respective risks. Early interventions, commonly performed within the first few months of life, can lead to increased complications associated with anesthesia, particularly in patients with comorbidities such as dextrocardia, which may involve structural heart defects (10). Children with cleft anomalies have been reported to exhibit a notable prevalence of associated congenital heart conditions, with studies indicating a significant percentage of patients presenting with cardiac anomalies, including dextrocardia (11).

Children with cleft lip and palate face significant challenges related to feeding, airway function, and facial growth due to the anatomical disruptions caused by these congenital anomalies. Feeding difficulties are prevalent among infants with cleft lip, particularly those with palatal involvement, as the gap between the oral and nasal cavities complicates the ability to generate suction during feeding, which can lead to ineffective breastfeeding and malnutrition (1,12).

Preoperative risk assessment for pediatrics patients with congenital anomalies, such as cleft lip and associated cardiovascular defects like dextrocardia, is crucial and should not only rely on genetic testing. A specific history and physical examination are essentials for the assessment process, allowing doctors to gather understandings about the patient's overall health, feeding habits, and any potential developmental delays (13). Basic investigations, including chest X-ray and echocardiography, are instrumental in ruling out complex congenital defects and identifying any anatomical variations or organ malposition that may be present, ensuring appropriate surgical planning (13,14). For example, a chest X-ray can detect dextrocardia and associated conditions like situs inversus, while echocardiography provides essential information about cardiac structure and function, allowing for comprehensive evaluation (14). Furthermore, monitoring growth and nutritional status is vital, particularly for patients with feeding difficulties, ensuring timely interventions to enhance growth and health outcomes (1). Therefore, a multifaceted approach that combines clinical evaluation with targeted imaging and growth monitoring is essential to adequately prepare for surgery and optimize patient outcomes.

Airway management in infants with cleft lip requires anticipating mask seal difficulty and potential intubation challenges (15). The positioning of the patient have a significant role during the induction of anesthesia; gentle handling is essential to maintain airway patency and prevent respiratory distress. For example, a semi-prone or lateral positioning can help prevent airway obstruction caused by the tongue falling into the cleft or obstructing the nasopharynx (16). Monitoring for potential airway complications during the perioperative period is critical, as studies have indicated that children born with cleft lip or palate have a higher risk of hospitalization due to airway infections and respiratory complications (17). Maintaining spontaneous respiration during induction and careful positioning can minimize risks (18). Intraoperatively, monitoring should include standard ECG lead placements, capnography, continuous pulse oximetry, and non-invasive blood pressure measurements, with an arterial line considered only in the presence of significant cardiac lesions or hemodynamic instability (19).

Avoiding rapid fluctuations in systemic vascular resistance is crucial in these patients, particularly given their cardiac considerations. Fluid management should consist of conservative isotonic boluses, closely monitored and adjusted based on hemodynamic metrics, avoiding agents that may induce significant myocardial depression, which is particularly relevant for patients with diminished cardiac reserves (20). Finally, patients should be observed in a high-dependency or neonatal intensive care unit setting for at least 24 hours postoperatively, particularly if any cardiac issues were present initially, to facilitate thorough monitoring and

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intervention as necessary (21). This comprehensive approach underscores the need for a multidisciplinary protocol in managing surgical patients with congenital anomalies and associated cardiac conditions to optimize safety and surgical outcomes.

In patients with cleft lip and dextrocardia, careful cardiac considerations are important due to the potential for conduction abnormalities and the unique anatomical challenges presented by this condition. Dextrocardia frequently involves different conduction patterns, where electrocardiograms may show atypical R-wave progression and P-wave inversions, complicating the interpretation of cardiac function (22). As such, preoperative hemodynamic monitoring strategies should be carefully planned, often requiring adjusted central line placements that account for the reversed cardiac anatomy (22). It is essential to avoid using anaesthetic agents that may compromise cardiac output; for instance, drugs such as volatile anaesthetics should be administered with caution, as they can lead to hemodynamic instability (23). Ensuring hemodynamic stability is crucial during surgery, particularly because children with dextrocardia can have pre-existing cardiac dysfunctions that increase the risk of perioperative complications (24,25). By carefully monitoring the heart and avoiding medications that could harm its function, anesthesiologists can help keep patients safer and achieve better results during surgery for this high-risk group.

In isolated dextrocardia with situs solitus, intravenous access is usually straightforward; however, clinicians should remain alert to potential vascular anomalies that may rarely accompany laterality defects (Dong et al., 2023; Bakalli et al., 2021). For example, using the right arm for venous access can sometimes be the simplest option, though the best site always depends on the patient's anatomy (Bakalli et al., 2021). Additionally, clinicians should be aware of the potential for vascular anomalies that can accompany dextrocardia, complicating catheter insertion (Dong et al., 2023). In these cases, continuous monitoring of heart function is important to make sure IV fluids and medications don't reduce cardiac output or worsen existing heart problems. Since patients with cleft lip and dextrocardia may also have feeding difficulties, it's especially important to quickly manage any IV complications, like infiltration or phlebitis, to provide the best care (Yuningsih et al., 2020; Birhane et al., 2017). In conclusion, careful planning of IV access and close monitoring are essential for safely managing patients with cleft lip and dextrocardia.

Choosing the right drugs and doses for patients with conditions like cleft lip and dextrocardia requires special attention to their unique anatomy and the possibility of reduced heart function. In dextrocardia, where the heart sits on the right side of the chest, anesthesiologists face added challenges such as interpreting ECG readings and placing catheters correctly. Placing ECG leads properly is especially

important to avoid false signs of ischemia during surgery, since this can greatly affect drug dosing and the overall anaesthetic plan (Fox et al., 2021; Subash et al., 2017).

A clear understanding of the heart's structure and any related defects is crucial, often requiring advanced imaging to guide preoperative planning and ensure anesthesia is given safely (Nakasone et al., 2021). Adjusting medication doses is especially important for patients with congenital anatomical differences and reduced heart function. In these cases, drug metabolism can be influenced by both the unusual physiology of dextrocardia and the limited cardiac reserve. Research shows that careful preoperative evaluation, including echocardiography, plays a key role in predicting potential complications and tailoring drug dosages appropriately (Chen et al., 2021).

Implementing individualized dosing strategies informed by comprehensive anatomical assessment can optimize therapeutic efficacy and reduce the risk of adverse effects, particularly when administering agents that influence cardiac contractility or rhythm, such as sedatives and anaesthetics. (Zeilmaker-Roest et al., 2018). This personalized strategy not only enhances perioperative safety but also supports more effective clinical outcomes by addressing the distinctive challenges posed by cleft lip and dextrocardia.

Managing patients with cleft lip and dextrocardia during surgery requires careful planning and continuous monitoring to address the unique anatomical challenges they present. Since the heart in dextrocardia is positioned on the right side of the chest, standard surgical positioning may not always work. Surgeons often adjust the setup to improve visibility and access, helping ensure the procedure is both safe and effective (26). In these cases, real-time heart monitoring is especially important to quickly detect issues like arrhythmias or low oxygen levels. Research shows that patients with congenital heart conditions face a much higher risk of developing arrhythmias during and after surgery. (23). The application of portable cardiac monitors provides the advantage of continuous tracking of heart rate and rhythm throughout the perioperative period. This ongoing assessment enables clinicians to promptly recognize sudden changes, such as arrhythmias or hemodynamic instability, and initiate timely interventions. By facilitating rapid detection and response, portable monitoring significantly enhances patient safety, particularly in individuals with underlying congenital anomalies or compromised cardiac reserve. (24).

In addition, providing supplemental oxygen and maintaining careful thermal regulation are essential aspects of intraoperative management in this case. Patients with cleft lip and dextrocardia may have reduced respiratory efficiency and impaired oxygenation, making supplemental oxygen crucial for sustaining adequate saturation and supporting favorable perioperative outcomes. (16). Maintaining normothermia is a critical component of intraoperative care, as hypothermia can worsen cardiovascular instability, especially during

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procedures involving prolonged aortic cross-clamp times. The use of active warming strategies helps prevent hypothermia and contributes to preserving both metabolic balance and hemodynamic stability throughout surgery (17). Collectively, these strategies underscore the importance of an integrative approach to intraoperative management, ensuring optimal care for patients with cleft lips and dextrocardia.

Cardiorespiratory monitoring in the recovery phase for patients undergoing cleft lip repair, especially those with complicating congenital anomalies like dextrocardia, is crucial to observe for potential respiratory or cardiac complications. Continuous monitoring of heart rate and respiratory rate can provide immediate feedback on the patient's recovery status, indicating any signs of cardiac decompensation or hypoxia that may arise (3,27). Given that patients with dextrocardia may already possess limited cardiac reserve, vigilant monitoring becomes essential to detect subtle signs of instability early and address them promptly. Indeed, early detection of deteriorating conditions allows healthcare providers to implement necessary interventions before they escalate into critical events, thereby enhancing patient safety and recovery outcomes.

In regard to postoperative care, early feeding strategies are particularly noteworthy in this patient population. Studies have shown that initiating feeding soon after cleft lip repair promotes faster recovery and reduces the risk of aspiration, which may be heightened in patients with underlying respiratory or cardiac issues (28). For infants and children, the assessment of feeding tolerance should be closely monitored, with attention to any signs of distress or discomfort that could signal cardiac decompensation or respiratory compromise. In patients with cleft lip and concurrent cardiac anomalies, the identification of these signs, such as increased respiratory effort or changes in heart rate, can guide clinical decisions in both feeding practices and overall recovery management (2). Therefore, a comprehensive approach integrating cardiorespiratory monitoring with early feeding protocols is essential to support optimal postoperative recovery for this vulnerable patient demographic.

In high-resource settings, perioperative evaluation of infants with craniofacial anomalies and dextrocardia often includes genetic testing and advanced cardiac imaging. However, in resource-limited environments, clinicians must rely on careful history-taking, thorough physical examination, echocardiography, and basic imaging to determine surgical readiness. The absence of major structural heart defects significantly alters the perioperative risk profile, permitting timely repair.

This article presents a case of a 3-month-old infant with unilateral cleft lip and isolated dextrocardia who underwent successful surgical repair. We combine the case description with a focused literature review to highlight practical

perioperative strategies for safe management in resource-limited contexts

CASE PRESENTATION

A 3-month-old male infant weighing 5.3 kg and measuring 59 cm in length presented with a complete right unilateral cleft lip. He was born at term by spontaneous vaginal delivery without perinatal complications. The family history was unremarkable. The infant showed feeding difficulty and mild undernutrition but no cyanosis, tachypnoea, or cardiac murmurs.

Preoperative vital signs were stable: heart rate 140 beats/min, respiratory rate 36/min, SpO₂ 98% on room air, and hemoglobin 11.2 g/dL. Transthoracic echocardiography confirmed dextrocardia with situs solitus, normal atrioventricular and ventriculoarterial concordance, preserved biventricular function (LVEF 66%), and no septal defects, patent ductus arteriosus, valvular abnormalities, coarctation, thrombus, or pericardial effusion. Abdominal ultrasound demonstrated situs solitus, consistent with isolated dextrocardia.

On general examination, the infant was alert and in moderate condition, with vital signs ranging from pulse 90–120 beats/min, respiratory rate 24–32/min, and body temperature 36–37 °C during hospitalization. Local examination revealed a right-sided cleft lip extending to the alveolar ridge, palate, and nasal structures, without signs of infection, oedema, or perioral complications.



Figure 1 Chest radiograph demonstrating right-sided cardiac apex consistent with dextrocardia.

Routine preoperative laboratory results were within normal limits, excluding infection or hematological contraindications. Preoperative consultations with pediatrics' and anesthesiology confirmed that the patient was fit for general anesthesia and reconstructive surgery. After multidisciplinary consultation with paediatrics, cardiology, anesthesiology, and plastic surgery, the team agreed to proceed with surgery following nutritional optimization.

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Written informed consent for surgery and case publication was obtained from the parents.

The surgical team performed primary cleft lip repair in August 2025 using the Fisher technique under general anesthesia. Anesthesiologist's adapted monitoring by reversing ECG lead placement to account for cardiac position. Induction was performed gently to maintain hemodynamic stability. The surgery proceeded uneventfully with negligible blood loss (estimated blood loss negligible; 10 gauze pads used), and no implants or specimens were required. Postoperatively, the patient was admitted for inpatient care, maintained in semi-Fowler's position, and did not require drains or nasogastric tubes. Oral amoxicillin (10–15 mg/kg every 8 h) and paracetamol (10–15 mg/kg every 8 h) were administered for infection prophylaxis and pain management. The infant recovered uneventfully with stable vital signs. The wound healed well, and the infant showed improved feeding efficiency and steady weight gain at follow-up.



Figure 2 Clinical photographs demonstrating the patient's preoperative cleft lip deformity and postoperative outcome following repair

DISCUSSION

This case illustrates that isolated dextrocardia does not substantially increase anaesthetic or surgical risk for cleft lip

repair when echocardiography rules out associated defects. The absence of intracardiac shunts, patent ductus arteriosus, or ventricular dysfunction allowed safe planning for early repair. In many low-resource hospitals, genetic testing and advanced imaging remain inaccessible. Clinicians can still provide safe care by integrating detailed clinical examination, chest radiography, echocardiography, and abdominal ultrasound. This pragmatic approach allows risk stratification without high-cost investigations.

In high-resource settings, standard guidelines for managing cleft lip repair in infants with dextrocardia often incorporate comprehensive genetic testing to identify underlying syndromes, such as Patau syndrome (trisomy 13) or Meckel-Gruber syndrome, which can inform tailored perioperative strategies and predict associated anomalies like congenital heart defects or laterality issues (6–9). For instance, genetic evaluation can reveal chromosomal abnormalities that correlate with a 45% prevalence of cleft lip in trisomy 13 cases, alongside cardiac malformations including dextrocardia, allowing for precise risk stratification and preoperative optimization (8,9). In contrast, in resource-limited settings, clinical acumen, detailed history, and basic diagnostics such as chest X-ray and echocardiography guide safe surgical planning. This pragmatic approach aligns with core principles of pediatric anesthesia guidelines while adapting to the absence of genetic testing (13,14). This adaptation maintains safety by focusing on functional assessments, such as evaluating cardiac structure and extracardiac anomalies, similar to how genetic data would guide interventions in well-equipped settings, but relies on empirical evidence from cross-sectional studies showing high correlations between cleft anomalies and heart defects (3,11). Ultimately, while genetic data enhances precision, the structured clinical pathway proposed here achieves comparable outcomes by integrating evidence-based adaptations from congenital cardiac management protocols (5,25,29).

Airway management in cleft lip patients poses predictable challenges, including mask seal difficulty and potential intubation difficulty. Dextrocardia itself does not alter airway anatomy. The anesthesia team maintained spontaneous ventilation during induction and carefully titrated anaesthetic agents to avoid hemodynamic swings. ECG leads were reversed to provide accurate rhythm monitoring. (6,7,21,22). For example, studies highlight that uncorrected cardiac malformations in dextrocardia significantly elevate mortality risk, and conditions like situs inversus could complicate vascular access or surgical positioning if not identified early (2,20,25,29). To mitigate these, a multifaceted preoperative evaluation is recommended, incorporating targeted imaging to detect situs variations and organ malposition, as demonstrated in case reports where echocardiography ruled out complex defects and guided safe interventions (13,14,22). Additionally, vigilant intraoperative monitoring with portable

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devices can address arrhythmias or reduced cardiac output, drawing from evidence in high-risk pediatric surgeries where such strategies reduced adverse events (23,24,26). By emphasizing nutritional monitoring and early feeding protocols to counteract malnutrition risks in cleft patients, this approach further minimizes complications like aspiration or growth delays, supported by systematic reviews on orofacial clefts (1,12,27).

Multidisciplinary collaboration strengthened the management plan. Paediatrics addressed nutritional optimization, pediatric cardiologist provided clearance, anesthesiologist prepared tailored monitoring and induction, and plastic surgery performed the repair. Such collaboration compensates for the lack of advanced diagnostics and maximizes safety. (3,5).. For instance, joint planning can adapt anesthesia protocols to avoid agents compromising cardiac function, while nursing input supports recovery through cardiorespiratory vigilance and early interventions (23,24). Such teamwork not only mitigates risks but also aligns with global guidelines for congenital anomalies, promoting safer, more effective management even without genetic insights (10,30,31).

LIMITATION

This is a single case, and genetic testing was unavailable. Subclinical syndromic associations cannot be fully excluded. Nevertheless, the case demonstrates how structured evaluation and pragmatic adaptation can yield safe outcomes in resource-constrained environments.

CONCLUSIONS

This case demonstrates that cleft lip repair can be performed safely in infants with isolated dextrocardia when a structured perioperative assessment confirms normal intracardiac anatomy. In the absence of genetic workup, clinicians can rely on thorough examination, basic imaging, and multidisciplinary coordination to guide decision-making. Active communication between specialties also the role of multidisciplinary coordination and careful anaesthetic adaptation allow good surgical and functional outcomes even in low-resource settings. Input from paediatric cardiology, anaesthesiology, paediatrics, and plastic surgery teams ensured that nutritional status, airway risks, and cardiac stability were jointly addressed. This collaborative approach is crucial in hospitals where advanced diagnostics or intensive care resources may be limited.

Previous literature has described higher anaesthetic risk in cleft patients with structural congenital heart disease, with increased incidence of perioperative desaturation and prolonged recovery. In contrast, isolated dextrocardia without intracardiac defects does not substantially elevate risk when basic monitoring and clinical precautions are observed. This reinforces the importance of distinguishing isolated dextrocardia from complex heterotaxy syndromes. As a single case, the report is not generalisable. Genetic workup

was unavailable, so subclinical syndromic associations cannot be entirely excluded. Nonetheless, the structured clinical evaluation and favourable surgical outcome suggest that safe cleft lip repair can be undertaken in similar contexts.

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