ISSN- O: 2458 - 868X, ISSN–P: 2458 – 8687
National Library of Medicine - ID: 101731606

International Journal of Medical Science and Innovative Research (IJMSIR)
IJMSIR : A Medical Publication Hub
Available Online at: www.ijmsir.com
Volume – 9, Issue – 6, November – 2024 , Page No. : 43 – 49

Perioperative Anesthetic Management of A Patient with Turner Syndrome Undergoing Two- Flap Palatoplasty Surgical Repair: A Case Report

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Citation this Article: Vishnu Datt, Sakshi Dhingra, Diksha Datt, Garima Sangwan, Shreya Khatri, Shivani Rathee, Shivani Dhaka, Shweta Chauhan, Sanjeev Kumar, "Perioperative Anesthetic Management of A Patient with Turner Syndrome Undergoing Two- Flap Palatoplasty Surgical Repair: A Case Report", IJMSIR - November - 2024, Vol – 9, Issue - 6, P. No. 43 – 49.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Turner's syndrome (TS) is one of the most common sex chromosomal abnormalities; monosomy (XO). The prevalence of TS is about 1 in 2000 to 1 in 2500 live female births. [1] Turner's syndrome can cause a variety of medical and developmental problems, including short height, webbed neck, broad chest with widely spaced nipples, low line hair, low-set ear, pubertal failure, infertility, and failure of ovaries to develop. TS may be associated with heart defects like bicuspid aortic valve (BAV), coarctation of the aorta (COA) and pulmonary arterial hypertension (PAH).[2] Such a combination of TS with cardiac and non-cardiac defects with a wide range of clinical presentations poses a challenge during perioperative management. TS with cleft palate can have a short trachea with a high location of the carina, risking endobronchial intubation and accidental extubation of the endotracheal tube. [3] The prevalence of cleft palate in patients with TS has not been described in the literature, as not many cases have been reported on the perioperative management of a patient with TS with cleft palate undergoing palatoplasty. We describe the

perioperative anaesthetic management of 14 years-6 months, short statured female with TS undergoing bardach,s two-flap palatoplasty repair after obtaining informed consent from the parents.

Keywords: Anaesthesia, Cleft palate, genetic, Intubations, palatoplasty, short trachea, Turner syndrome

Introduction

Turner syndrome is a genetic disorder characterised by an abnormal X chromosome and the female is 45X karyotype. It occurs in 1 in 2000 to 2500 live female births. The clinical picture may be highly variable including a short stature, webbed neck, maxillary and mandibular hypoplasia, short trachea and high location of the carina and absence of the menarche. TS may be associated with several cardiac and non-cardiac lesions like BAV, COA, PAH, and hypothyroidism. TS patients with these associated lesions with a wide clinical spectrum pose a unique challenge during perioperative anaesthetic management and are at increased risk of hemodynamic instability, and morbidity and mortality. This report of a rare case illustrates the perioperative anaesthetic management in a 14yrs-6-month-old patient of TS with incomplete cleft palate undergoing two flap palatoplasty.

Case Report

14 yrs -6 months-female presented with incomplete bilateral cleft palate scheduled for Bardach's two- flap palatoplasty surgical repair. The clinical history was non-significant except for the delayed milestones. On examination, she was short-statured (125 cm), weighing 20 kg, showed that the patient was at less than 3rd percentile, and so BMI of less than 3rd percentile with respect to the 14yrs-6 months female using an Indian Academy of Paediatrics (IAP) growth chart. She has facial dysmorphism; concomitant squint, increased intercanthal distance, webbed neck, long philtrum of the nose with nasal septum depression, low placed ears. In addition, fissured bifid tongue, receding hairline, and broad chest with widely spaced nipples. [Figure 1a &b], [Figure 2] She has not yet reached to her menarche. This whole spectrum of presentation is suggestive of turner's syndrome. Furthermore, she has bilateral incomplete cleft palate and missing multiple teeth.[Figure 3a & b] Auscultation of chest and heart revealed normal air entry, and normal S1 and S2 heart sounds. Her various haematological and biochemical values were within the normal limits. Chest X-Ray revealed a shorter trachea length as per her age (8.51cm vs 11.2cm) with a higher location of the bifurcation(carina). [Figure 4] ECG showed non-specific widespread T- wave abnormalities. 2-D echocardiography evaluation revealed good biventricular functions and ruled out the presence of any cardiac anomaly. Ultrasonography study for abdomen and pelvic organs was unremarkable. However, ovarian parenchymal like structures were noted in the bilateral ovaries. Though, further assessment of the ovaries, karyotyping, hormonal assay could not be performed due to limited financial support. Finally on the bases of the clinical presentation and physical examination the diagnosis of the TS with incomplete bilateral cleft palate was established. After obtaining the detail informed consent, she was posted for the cleft palate repair under the smile train scheme.

In The OR, standard ASA monitoring (ECG, SPO2, NIBP, capnography and temperature) was attached. The Premedication was given with injection glycopyrrolate (0.2mg) and midazolam (1mg). A balanced general anaesthesia induction was performed using propofol (40mg), fentanyl(40mcg), sevoflurane (1%) in oxygen (FIO2- 0.5) and atracurium citrate (10mg) was used to facilitate endotracheal intubation with south-facing RAE, 5.5 mm.ID, endotracheal cuff tube, fixed at 14 cm mark.

Throat was packed with wet roll gauze. An airway pressure of 17 mmHg, and EtCO2 of 30-37 mmHg were observed throughout the surgery. Anaesthesia was maintained with sevoflurane (1-2%), intermittent fentanyl, midazolam, and paracetamol(300mg) was also used as an analgesic adjuvant. Total blood loss was approximately 200cc. Bilateral grater palatine nerve block was given with 0.25%, 0.5ml of bupivacaine for postoperative analgesia. Surgery lasted for 2 hours, and neuromuscular block was reversed with a mixture of neostigmine 1 mg and glycopyrrolate 0.2mg. Throat pack was removed after confirmation of the adequate haemostasis. On tracheal extubation she was fully conscious and a SPO2 of 99-100% was maintained on air. Patient was shifted to recovery room for observation in right lateral position and rest of the course was uneventful.

Discussion

Turner syndrome (TS) is a genetic disorder characterised by an abnormal X chromosome and the female is 45X karyotype. [1,2] TS affects approximately 1 in 2000 to 1 in 2500 live female births.[1,2,4] Table 1 enlisted the characteristic clinical features of the TS.[2,3,5] This patient of TS also had typical features and an isolated incomplete cleft palate without any other associated congenital anomalies.(Figures-1a&b, 2,3a). Patients of TS also have elevated levels of follicle-stimulating hormone.[6]

Nowadays hormonal treatment with recombinant human growth hormone (HGH) is the most recommended therapeutic approach in the TS, which significantly improves the quality of life. HGH can help to normalize skeletal growth when properly administered at an early age, at approximately 9 months of age.[7] International guidelines of 2016 have recommended initiating GH therapy early at 4–6 years of age, and preferably before 12–13 years of age, if the child already has evidence of growth failure, using a GH dose of 45–50 µg/kg per day increasing to 68 µg/kg per day if actual height potential is substantially compromised, and therefore, early diagnosis is important.[4]

Patients with TS have an increased risk of cardiac defects like BAV, COA, partial anomalous pulmonary venous connections (PAPVC), elongated transverse aortic arch, and PAH.[8] The incidence of cardiac defects in TS is approximately 50% and are the major contributors to early death. Therefore, early diagnosis of COA is important to prevent serious complications such as infective endocarditis, aortic aneurysms, aortic dissection, and aortic dilatation and heart failure and cardiac conduction defects.[9]. Approximately 50% of Patients with TS have renal anomalies like collecting system malformation, positional abnormalities and horseshoe kidney predisposing to the urinary tract infection. [10] The autoimmune disease like hypothyroidism may also be associated with TS, that increases the risk of development of cardiovascular instability due to decreased intravascular volume and preload, and blunted baroreceptor response, and decreased cardiac output, and makes the anaesthetic management more complicated.[11] Though this patient did not have any major cardiac and non-cardiac associated anomalies, however, it is worth to describe that these associated defects pose a challenge during perioperative management and increase morbidity and mortality.

Generally, cleft palate is repaired between 3 and 9 months to promote normal speech development and reduce nasal regurgitation. The American Cleft Palate-Craniofacial Association (ACPA) recommends palatoplasty prior to 18 months.[12]These patients with cleft palate may exhibit with frequent URTI with chronic

rhinorrhoea due to food reflux into the nasal passages and may require antibiotic therapy to prevent the postoperative respiratory complications.[13] Delayed in the repair have been reported where access to medical care is difficult, or as a result of lack of awareness and knowledge of the importance of the early surgery. Primary repair for the adult patient is different from the infants because adults have completed the facial growth.[14] The delayed palatoplasty repair like in this patient may interfere less with the midfacial growth, but speech development is adversely affected. Primary palatoplasty is more difficult in adults, because in adults cleft palate attains wider, and the transverse palatal surface around the transverse palatine sutures is sharp, and palatal segments are more vertically displaced due to location of the tongue between the cleft palate for cleft closure. Therefore, the incidence of oronasal fistula after palatoplasty is higher.[14]

The described patient was managed under the smile train scheme. The Smile Train is the world's largest cleft-focused organization, that empowers local medical professionals with the training, funding, and resources to provide **100%-free cleft surgery globally**.[15]

Generally, a balanced anaesthesia technique should be practiced aiming an enhanced recovery by choosing shorter-acting premedication (midazolam, glycopyrrolate), volatile anesthetics, (sevoflurane 1 MAC), intravenous anaesthetics (propofol 2mg/kg, or etomidate 0.2-0.3 mg/kg), lower doses of opioids (fentanyl 1-2 mcg/kg). The endotracheal intubation should be performed with oral south pole, Ring – Adair – Elwyn (RAE) tracheal tube, and fixed in the midline below the lower lip to allow an optimal surgical access and to prevent the obstruction of the tube by mouth gag when it is positioned. The throat should be packed with a wet roll gauze to avoid the aspiration of the blood and other debris, and to support the tube position in midline. The air entry should be rechecked after throat pack, as insertion or removal of the throat pack can lead to accidental extubation of the tube. These patients should be intubated with smaller size tube and needs to be fixed at lesser length as compared to the healthy females of the same age, considering the shorter length and small diameter of the trachea, as well as the higher location of the carina, These can predispose to endobronchial intubation particularly with the South-facing RAE tube with long preform Knee, and accidental extubation when the tracheal is under traction.[3] A standard ASA monitoring (pulse oximetry, ECG, non-invasive arterial BP, capnography, and temperature) is sufficient, During surgery, the focus should be to attenuate the stress, and pain response, fast track recovery, and maintain euvolemia, normothermia, hemodynamic stability, and to avoid endobronchial intubation and accidental extubation and even tube compression.

Postoperative care involves often a lateral or semi-prone position to allow drainage of any secretions. administering prophylactic antiemetics such as Injection hydrocortisone(5mg/kg) and ondansetron (0.1 mg kg^{-1}). Multimodal analgesia for cleft procedures includes fentanyl (0.25-0.5 mcg/kg boluses), or diclofenac(2mg/kg) or paracetamol(20mg/kg), and dexmedetomidine (0.3-0.5 mcg/kg/hr) infusion with $\alpha_2$agonist actions, has been used for arousable sedation, analgesia, anti-inflammatory and control of shivering. Other options include, NSAIDs (e.g. ibuprofen 5–10 mg /kg or ketorolac 0.5 mg /kg i.v.), ketamine (0.1–0.2 mg/ kg. i.v.), Furthermore regional anaesthesia using a greater/ lesser palatine and nasopalatine nerve blocks have been used successfully for cleft palate. [16,17]

Conclusion

TS is usually associated with facial dysmorphism, webbed neck, low placed ears, increased intercanthal distance, receding hairline, and cubitus valgus and absence of menarche in a short statured female. Some patients with rural and poor socioeconomic background may present with delayed cleft repair due to difficult access to medical care, or lack of awareness and knowledge of the importance of the early surgery. Primary repair for the adult patient is different from the infants because adults have completed the facial growth and adversely affects the speech development. The patients of TS with isolated non- syndromic cleft palate for palatoplasty can be managed under balanced GA technique, vigilant postoperative care, and multimodal pain and vomiting control.[18]

References

1. Cui X, Cui Y, Shi L, Luan J, Zhou X, Han J. A basic understanding of Turner syndrome: Incidence, complications, diagnosis, and treatment. Intractable Rare Dis Res. 2018 Nov;7:223-228
2. Bondy CA., Turner Syndrome Study Group. Care of girls and women with Turner syndrome: a guideline of the Turner Syndrome Study Group. J Clin Endocrinol Metab. 2007 ;92:10-25.
3. Maranhão MV. Turner syndrome and anesthesia. Rev Bras Anestesiol. 2008 Jan- Feb;58 :84-9.
4. Gravholt CH, Andersen NH, Conway GS, Dekkers OM, Geffner ME, Klein KO, et al. International Turner Syndrome Consensus Group. Clinical practice guidelines for the care of girls and women with Turner syndrome: proceedings from the 2016 Cincinnati International Turner Syndrome Meeting. Eur J Endocrinol. 2017 Sep;177: G1-G70.
5. Apperley L, Das U, Ramakrishnan R, Dharmaraj P, Blair J, Didi M, et al. Mode of clinical presentation and delayed diagnosis of Turner syndrome: a single Centre UK study. Int J Pediatr Endocrinol. 2018;2018:4.
6. Lunding SA, Aksglaede L, Anderson RA, Main KM, Juul A, Hagen CP,et al. AMH as Predictor of Premature Ovarian Insufficiency: A Longitudinal Study of 120 Turner Syndrome Patients. J Clin Endocrinol Metab. 2015 Jul;100: E1030-8
7. Tommaso Aversa, Alessandra Li Pomi, Giorgia Pepe, Domenico Corica, Maria Francesca Messina, Roberto Coco, et al. Growth Hormone treatment to final Height in Turner Syndrome: systematic Review. Clinical Therapeutics. 2024; volume 46, Issue 2: 146-153
8. Noordman ID, Fejzic Z, Bos M, Duijnhouwer AL, Weijers G, Kempers M, et al .Cardiac abnormalities in girls with Turner syndrome: ECG abnormalities, myocardial strain imaging, and karyotype-phenotype associations. Am J Med Genet A. 2021 Aug;185 :2399-2408.
9. Claus H Gravholt, Niels H Andersen, sophie Christin-Maitre, Shanlee M Davis et al. The international Turner Syndrome Consensus Group, Philippe F Backeljauw, Clinical Practice guidelines for the care of girls and women with Turner syndrome: Proceedings from the 2023 Aarthus International Turner syndrome meeting. European Journal Of Endocrinology, Volume 190, issue 6, June 2024, Pages G53-G151
10. Berglund A, Viuff MH, Skakkebæk A, Chang S, Stochholm K, & Gravholt CH. Changes in the cohort composition of turner syndrome and severe non-diagnosis of Klinefelter, 47,XXX and 47,XYY

syndrome: A nationwide cohort study. Orphanet Journal of Rare Diseases. 2019; 14, 16. 10.1186/s13023-018-0976-2

11. Sudha P, Koshy RC, Pillai VS. Undetected hypothyroidism and unexpected anesthetic complications. J Anaesthesiol Clin Pharmacol.2012 Apr; 28: 276-7
12. Shaffer A.D., Ford M.D., Losee J.E., Goldstein J., Costello B.J., Grunwaldt L.J., Jabbour N. The Association Between Age at Palatoplasty and Speech and Language Outcomes in Children With Cleft Palate: An Observational Chart Review Study. Cleft Palate Craniofac. J. 2020; 57:148–60
13. Shaw W, Semb G, Lohmander A, et al. Timing of Primary Surgery for cleft palate (TOPS): protocol for a randomised trial of palate surgery at 6 months versus 12 months of age. BMJ Open 2019; 9(7):e029780
14. Lee JS, Lee JY, Kim YD, Song JM, Choi WH. Primary repair of untreated cleft palate in an elderly patient: a case report. J Korean Assoc Oral Maxillofac Surg 2021;47:47-50.
15. Singh SK. Smile Train: The ascendancy of cleft care in India. Indian J Plast Surg. 2009 Oct;42 Suppl(Suppl):S192-8.
16. Vishnu Datt, Diksha Datt, Isha Singh, Priyanka Dhahiya, Saket Agarwal, Ashtha Gaba,et al . Perioperative Anaesthetic Management for an Isolated or Combined Tetralogy of Fallot (TOF) and Cleft Palate/Lip Repairs - A Systematic Review and Meta-Analysis‖, IJMSIR - July - 2024, Vol – 9, Issue - 4, P. No. 59 -79
17. Denning S, Ng E, Wong Riff KWY. Anaesthesia for cleft lip and palate surgery. BJA Educ. 2021 Oct;21(10):384-389.
18. Suleiman NN, Luedi MM, Joshi G, et al. Perioperative pain management for cleft palate surgery: a systematic review and procedure- specific postoperative pain management (PROSPECT) recommendations Reg Anesth Pain Med 2023;0:1–7.

Legends Table and Figures

Short stature,	Cubitus valgus,
Webbed neck	Multi-pigmented naevi,
Foetal lymphedema,	Broad chest
Facial dysmorphism,	Widely spaced nipples,
Drooping eye lids (ptosis)	Absence of menarche as per her age.
Squint,	Small and receding lower jaw;
Long philtrum of the nose	Narrow palate
Nasal septum depression,	A missing knuckle in a digit (finger or toe),
Low placed ears, elongated ears, cup-shaped ears and thick ear lobes	making that digit shorter than the rest.
	Flat Feet (pes planus)
Increased intercanthal distance, Receding hairline	Chronic Middle ear infection leading to hearing loss

Table 1: Enlisted characteristic clinical features of the Turner syndrome. [2,3,5]

Figure 1 A & B: Preoperative picture of the patient with front(a) and lateral(b) face position shows a concomitant squint, nasal septum depression, increased intercanthal space, low placed ears, receding hairline and short statured female. All features suggestive of Turner syndrome.

Figure 2: The intraoperative photograph revealed that the trachea being intubated with south pole cuffed endotracheal tube. In addition, a broad chest with widely spaced nipples can also be observed, another important feature of the turner syndrome. Also, the electrodes are attached for the ECG monitoring.

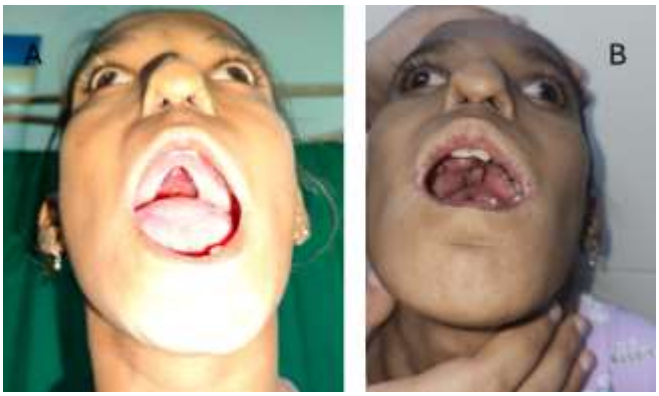

Figure 3 A&B: The preoperative(a) and postoperative(b) examination of the oral cavity revealed an incomplete bilateral cleft palate. In addition, several missing teeth and normal lips without any cleft. (b) photograph of the open mouth of the same patient confirms the adequate two flap palatoplasty repair

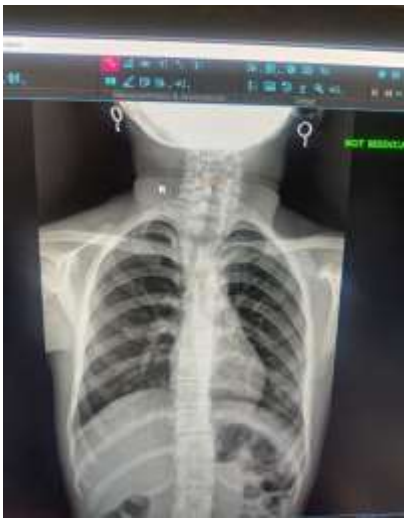

Figure 4: Chest X-Ray (PA view) shows a normal cardiac size along with clear lung fields and clear outline chest cavity. However, it depicts a small trachea with a size of 8.5cm and higher located carina, increasing the risk of both the endotracheal intubation or accidental extubation with the neck movements during cleft palate repair.