Management of congenital and acquired airway pathologies in newborns by a cross-disciplinary committee at a third level hospital

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Abstract

Introduction. Neonatal airway examination through flexible/rigid bronchoscopy has proved to be useful in the presence of persistent stridor and extubation failure, as well as to assess complications following cardiac surgery. At our institution, these examinations are carried out by a pulmonologist, a neonatologist, an otorhinolaryngologist, and a pediatric surgeon from the pediatric airway committee, established in 2014.

Objective. To analyze the airway examinations performed in neonates during their stay at the neonatology/neonatal intensive care unit since the airway committee was established.

Materials and methods. A retrospective study of the airway examinations conducted in neonates from 2015 to 2019 was carried out. Clinical and demographic data, number of examinations, indications, findings, and complications were collected. Results are presented as mean and standard deviation. Statistical significance was established at $p < 0.05$.

Results. 92 airway examinations were analyzed in 51 patients (54.9% of whom were female). 51% of the patients were premature. Extubation failure and persistent respiratory symptoms following successful extubation were the most frequent indications for airway examination (35.3%). Stratification by gestational age or weight at birth was not associated with an increased risk of pathological findings at examination ($p > 0.05$). The most frequent finding was vocal cord paralysis (n = 14; 27.5%). In 10 patients (19.6%), no pathological findings were observed.

Conclusion. Airway examination is useful in patients with stridor to identify vocal cord paralysis following extubation failure. It also allows congenital airway pathologies to be diagnosed and treated. The number of examinations with no pathological findings was similar to that reported in international series.

Key Words: Airway extubation; Newborn; Diagnosis; Rigid bronchoscopy; Flexible bronchoscopy.

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Manejo de la patología congénita y adquirida de la vía aérea en el neonato por parte de un comité multidisciplinar en un centro terciario

Resumen

Introducción. La exploración de vía aérea mediante broncoscopia flexible/rígida en el neonato ha demostrado utilidad en el estridor persistente, extubaciones fallidas o para valorar complicaciones tras cirugía cardiaca. En nuestro hospital estas exploraciones son practicadas por un neumólogo, neonatólogo, otorrinolaringólogo y cirujano pediátrico del Comité de Vía Aérea Pediátrica, formado en 2014.

Objetivo. Analizar las exploraciones de vía aérea practicadas a neonatos durante su estancia en Neonatología/Unidad de Cuidados Intensivos Neonatales desde la constitución del Comité de Vía Aérea.

Materiales. Estudio retrospectivo de exploraciones de vía aérea practicadas a neonatos de 2015-2019. Se recogen datos clínicos y demográficos, número de exploraciones, indicación, hallazgos y complicaciones. Se presentan las medias con su desviación estándar. Se consideró un resultado estadísticamente significativo cuando $p < 0,05$.

Resultados. Se analizaron 92 exploraciones de vía aérea en 51 pacientes (género femenino: 54,9%). El 51% de los pacientes fueron prematuros. La extubación fallida y la persistencia de sintomatología respiratoria tras una extubación satisfactoria fueron las indicaciones más frecuentes (35,3%). La estratificación por edad gestacional o por peso al nacimiento no se asociaba a un mayor riesgo de presentar hallazgos patológicos en la exploración. El hallazgo más frecuente fue la parálisis de cuerda vocal (n = 14; 27,5%). En 10 pacientes (19,6%) no se encontraron hallazgos patológicos.

Conclusión. La exploración de la vía aérea es útil en pacientes con estridor postextubación y para identificar parálisis de cuerda vocal tras extubación fallida. Además, permite el diagnóstico y tratamiento de patologías congénitas de la vía aérea.

Palabras Clave: Extubación de la vía aérea; Recién nacido; Diagnóstico; Broncoscopia rígida; Broncoscopia flexible.

INTRODUCTION

Airway examination using a flexible bronchoscope (FB) or a rigid bronchoscope (RB) has demonstrated to be useful in the diagnostic and therapeutic management of newborns with suspected congenital and acquired airway abnormalities. Bronchoscopy provides a well-defined anatomical detail, allows airway structure and mobility from the nasal cavity to the bronchial tree to be observed, and demonstrates potential intrinsic or extrinsic compressions[1-5]. Failed extubation following surgery or as a result of long-term invasive mechanical ventilation is one of the main indications of airway examination. Extubation failure, which is defined as the need for reintubation in the first 48 hours following the first extubation attempt, increases ICU stay and patient morbidity. Reported incidence of failed extubation ranges from 22% in full-term newborns to up to 40% in extremely premature newborns[4,5].

In addition, airway examination is useful in patients with stridor, choking, or hoarse cry as a result of suspected mechanical airway disorder or lack of movement of any of its components[4]. It also allows mechanical complications following cardiac and cervical surgery to be diagnosed[6,7].

Airway examination may be carried out at the crib side at the neonatal intensive care unit –without moving the patient to the operating room– but under conscious sedation and without requiring the presence of a pediatric anesthesiologist[3,4]. This is a safe technique both in newborns and in pediatric patients[5], with an incidence of major complications (massive bleeding or pneumothorax) below 5%. Most complications are related to transbronchial lung biopsies, which occur primarily in the adult population, with a 0.04% mortality rate[1].

Specific airway units staffed by cross-disciplinary teams made up of neonatologists, intensivists, pulmonologists, anesthetists, otorhinolaryngologists, voice therapists, maxillofacial surgeons, and pediatric surgeons have been created worldwide. According to Torre et al., these highly specialized cross-disciplinary teams have achieved better surgical results in these patients[6].

OBJECTIVE

To assess and analyze the diagnoses and indications of the neonatal airway examinations carried out by the members of the Pediatric Airway Committee (PAC) from our hospital since it was established (study period: 2015-2020).

To estimate the proportion of examinations without pathological findings in our neonatal care unit.

To find out whether prematurity and low weight at birth are associated with pathological examination.

To describe the most relevant clinical cases from our series.

MATERIALS AND METHODS

Study type

A retrospective study of the newborns admitted at the Neonatology Department or at the Neonatal Intensive Care Unit with symptoms associated with congenital or acquired airway abnormalities, and undergoing FB and/or RB, from 2015 –when the PAC was established– to 2020 was carried out.

Clinical variables collected included sex, gestational age (weeks, days), weight at birth, indication for airway examination, type and number of examinations, examination findings, diagnosis following examination, and perioperative complications. Patients were classified into premature newborns (< 37 gestation weeks) and full-term newborns (≥ 37 gestation weeks).

Quantitative variables were expressed as mean and standard deviation, whereas qualitative variables were expressed as absolute frequency and percentage**.**

Pearson's chi-squared test (X^2) was used to analyze differences in qualitative variable distribution. Student's T test for unpaired data was used to analyze quantitative variables. Statistical significance was established at $p < 0.05$.

Exclusion criteria

Newborns with suspected esophageal atresia (of any type) undergoing FB in the perioperative period to identify the presence of tracheoesophageal fistula were excluded.

Technique

Fibrobronchoscopy

The patient, under conscious sedation, is placed on a supine position with an adequately sized roller underneath their shoulders. The fibrobronchoscope is introduced through the nostril or the mouth until the epiglottis and the mobility of the vocal cords are visualized. Examination is pursued with the patient under spontaneous breathing, while assessing the subglottal region, the trachea, and the tracheal rings down to the carina. The right main bronchus and its segmental bronchi are explored. The left bronchial tree is then examined. In case the patient has bronchial reactivity, 1 ml of 1% lidocaine may be instilled at the level of the vocal cords. This dose can be repeated (with a maximum of 2 ml) in patients with persistent bronchial reactivity by instilling 1% lidocaine at the level of the carina. In some cases where laryngeal or vocal cord assessment is not required, the airway is examined by means of an endotracheal tube (ETT), without the need for lidocaine instillation.

Rigid bronchoscopy

The patient, under general anesthesia and full relaxation, is placed in a supine position with cervical hyper-

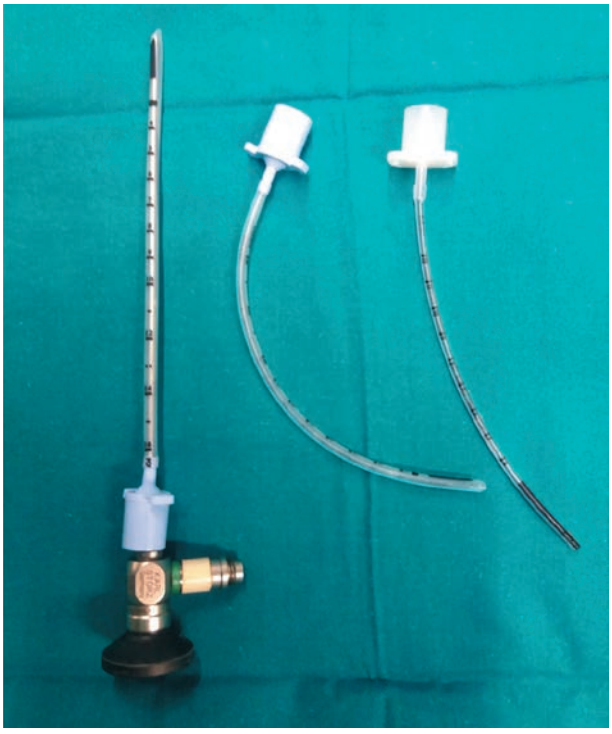

Figure 1. Instruments used for airway assessment and dilatation: 2 mm Hopkins 0º scope (useful length: 18 cm) and endotracheal tube of various sizes, which allows for progressive dilatation. The picture features an ETT without balloon, with the following sizes, from left to right: 3 mm, 2.5 mm, and 2 mm.

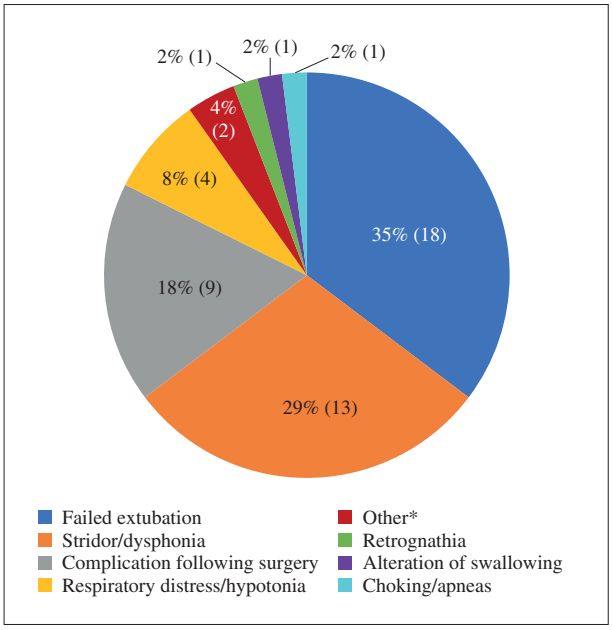

Figure 2. Distribution of indications for airway examination at our unit (percentage, in brackets: number of patients). *Others: airway assessment in a patient with gastroparesis and in a patient with Moebius syndrome.

extension and an adequately sized roller underneath their shoulders. The appropriate laryngoscope is placed from the right side of the oral cavity while mobilizing the tongue to the left. The epiglottis is then visualized and lifted, while instilling 1% lidocaine into the vocal cords under direct vision –unless a previous FB has been carried out. A *Hopkins 0º* scope–2-4 mm diameters, according to gestational age and weight –is introduced so as to assess the larynx, the mobility and structure of the vocal cords, and the caliber of the subglottal region and the trachea until the carina and both bronchi are visualized.

In some cases, as in the presence of non-congenital subglottal stenosis –defined as a subglottis with a < 4.5 mm diameter in full-term newborns, and with a < 3.5 mm diameter in premature newborns[4]–, dilatation may be carried out in the same procedure using an ETT, while progressively increasing its size until an adequate diameter is achieved according to patient age and weight. Figure 1 shows the instruments used for airway visualization and dilatation. Once the stenosis has been adequately identified, the ETT is placed within the scope, and under direct vision, both devices are introduced until the stenosis has been reached. The scope is then removed, and the ETT is left in place for 1-5 minutes) before removal. A final review is carried out to verify whether the stenosis has been adequately dilated using the *Hopkins 0º* scope. Under direct vision, topical corticosteroid is applied at the level of the dilated area and the vocal cords using a swab.

RESULTS

Data from 136 airway examinations in 75 newborns were collected. 24 patients (44 examinations) with esophageal atresia undergoing pre-surgery airway examination as a perioperative protocol were excluded.

The remaining 51 patients (female patients: 54.9%; male patients: 45.1%) were analyzed, with a total of 92 airway examinations. Mean gestational age was 33 ± 6 weeks. 51% of the patients were premature. Mean weight was 2.102 ± 1.166 g.

Indications for airway examination and airway examination findings are featured in figure 2 and table I. Failed extubation and persistent respiratory symptoms following successful extubation were the most frequent indications (35.3%). The most frequent finding was vocal cord paralysis (n = 14; 27.5%). In 10 (19.6%) patients, no pathological findings were found at airway examination.

Stratification by gestational age or weight at birth was not associated with a greater risk of pathological findings at examination (p = 0.982 and p = 0.627, respectively).

The most commonly used technique was FB, which was performed as a single test without further examinations in 80.4% of the cases.

Table I. Diagnosis following airway examination.

Examination findings	*n (%)*
Normal	10 (19.6)
Vocal cord paralysis	14 (27.5)
Airway edema	12 (23.5)
Laryngomalacia	6 (11.8)
Tracheomalacia	4 (7.9)
Airway obstruction	3 (5.9)
Subglottal stenosis	1 (1.9)
Tracheal stenosis	1 (1.9)

Airway obstruction caused by the clip used for the closure of persistent ductus arteriosus in one patient, and by vocal cord granuloma following intu-bation in the other two patients.

We present five cases we believe could be relevant owing to how rare they are and/or how complex management is:

- **Patient 1.** 20-day-old male newborn with history of hydrocephalus as a result of bilateral intraventricular bleeding and placement of a ventriculoperitoneal shunt valve. Airway examination was conducted owing to the presence of respiratory stress and predominantly expiratory stridor 24 hours following extubation. Membranous stenosis at the level of the subglottis was found. It was dilated in the same procedure using a rigid bronchoscope.
- **Patient 2.** 20-day-old female newborn diagnosed with Moebius syndrome and cutaneous capillary malformations. FB did not show any malformations of the airway mucosa.
- **Patient 3.** 17-day-old male newborn with tetralogy of Fallot and tracheal compression as a result of a double aortic arch. Airway examination demonstrated the presence of tracheal stenosis. A 6 x 20 mm Dumon® silicone tracheal stent (Novatech, France) was placed, and the patient could be extubated following surgery.
- **Patient 4.** Female patient diagnosed with persistent ductus arteriosus and pulmonary bleeding. She underwent ductus arteriosus surgery when she was 12 days old. Respiratory stress with left pulmonary atelectasis was noted following extubation. Given the lack of improvement, decision was made to examine the airway using a FB, which showed an entirely collapsed left main bronchus. CT-scan confirmed extrinsic compression caused by a clip at the level of the bronchus (Fig. 3). The clip was urgently removed through thoracotomy. Patient hemodynamic status improved following surgery.
- **Patient 5.** 5-day-old male neonate born at gestation week 37 with suspected tracheoesophageal fistula as a result of coughing and choking following breastfeeding. Airway examination revealed a type III laryngotracheoesophageal cleft (Fig. 4), which was surgically repaired perinatally.

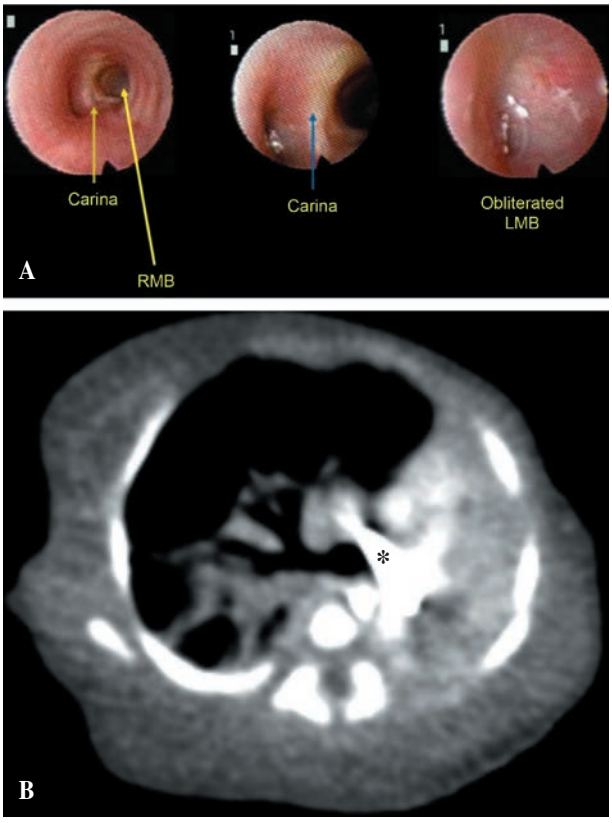

Figure 3. A) Left picture: FB demonstrating the trachea and the origin of the right main bronchus (RMB). Center picture: carina, RMB (partial image), and left main bronchus (LMB) without lumen. The right picture shows entire LMB collapse through the clip at the level of the ductus arteriosus. B) CT-scan demonstrating complete atelectasis of the left lung and presence of a clip obliterating the lumen of the left main bronchus at the level of the ductus (asterisk).

DISCUSSION

RB was used as a therapeutic option in pediatric patients for foreign body removal purposes before the diagnostic use of FB was first described in 1978[8]. Since then, airway examination –both using RB and FB– has played a key role in the diagnostic and therapeutic management of pediatric patients with airway pathologies, either acquired or congenital[8].

In the last years, the low incidence of these pathologies, their diagnostic complexity, and the fact these patients need to be managed by a specialized group has led to the creation of highly complex cross-disciplinary units[6-9].

According to Torre et al. (2011) and Kocyildirim et al. (2004), cross-disciplinary teams should be created at third-

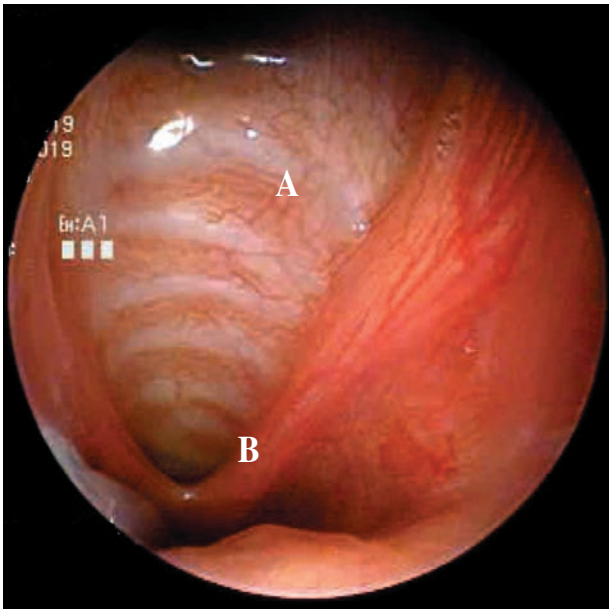

Figure 4. Airway examination of a newborn with type III laryngotracheoesophageal cleft. The picture features: A) incomplete tracheal rings in its posterior portion; B) tracheoesophageal party wall.

level institutions specialized in airway pathologies, since they allow for better surgical results and reduce patient management costs[6,7]. Following their guidelines, airway examinations at our healthcare facility are carried out by PAC members since the PAC was established in 2014 –it came into operation one year later.

Nowadays, the PAC consists of a pediatric cross-disciplinary team made up of neonatologists, pulmonologists, otorhinolaryngologists, anesthetists, maxillofacial surgeons, and pediatric surgeons. In addition, the fact Neonatal Intensive Care Unit members are in direct contact with the PAC allows studies to be scheduled in a very short time in patients with respiratory symptoms (stridor, hoarse cry, cyanosis, and/or desaturation) or suspected airway disorders.

In our series, the incidence of post-intubation subglottal stenosis was 1.9%, similar to that described in the literature, and associated with long-term intubation in premature patients[9]. Endoscopy remains the technique of choice in the post-intubation diagnosis of subglottal stenosis, and it allows it to be treated through dilatation[6,10]. Even though dilatation can be achieved using balloons, our institution advocates the use of increasingly large ETTs to allow for a nice circumferential dilatation. In our series, no complications associated with ETT dilatation, such as tracheal/bronchial rupture or trauma, were found, contrarily to the literature[8]. This allows for progressive dilatation without the need for the patient to be apneic. Even though they typically appear at the age of 2 months, laryngomalacia (LM) and tracheomalacia (TM) are two frequent diagnoses in newborns with refractory respiratory distress, wheezing, or cyanosis. In some cases, this may cause potentially life-threatening events. In our series, the LM/TM finding was reported in 6 and 4 patients, respectively. Various diagnostic tools, such as fluoroscopy and contrast-enhanced CT-scan[10], have been proposed for LM/TM, but direct visualization of the airway's dynamic narrowing using endoscopic examination remains the technique of choice for diagnostic purposes[11].

Direct lesion of the vocal cords may occur in pediatric cardiovascular surgery when closing the ductus arteriosus and the ductus venosus[12-14] as a result of contusion or lesion of the recurrent laryngeal nerve[12], usually unilateral[14], and typically in premature patients with extremely low weight[12]. Vocal cord lesion may also occur following intubation in patients undergoing any type of surgery, since it may give rise to granuloma and stenosis, which reduce vocal cord mobility[13]. Shen et al. suggested bilateral vocal cord paralysis has an idiopathic cause[14]. In our series, vocal cord lesion was observed in 27.5% of the patients –we believe most of them were secondary to long-term intubation.

Given the low incidence of congenital and acquired airway pathologies, as well as the characteristics of newborns, a cross-disciplinary team at a third-level hospital is required for patient management purposes, as it is the case with our airway unit.

Airway examination using FB or RB is useful and safe in newborns following failed extubation, primarily as a result of subglottal stenosis. It also allows vocal cord paralysis following surgery to be diagnosed, and it has been demonstrated as a useful technique to examine newborns with airway malformations.

REFERENCES

1. Miller R, Casal R, Lazarus D, Ost D, Eapen G. Flexible bronchoscopy. Clin Chest Med. 2018; 39: 1-16.
2. Francom C, Best C, Eaton R, Pepper V, Onwuka A, Breuer C, et al. Clinical validation and reproductibility of endoscopic airway measurement in Pediatric aerodigestive evalutaion. Int J Pediatr Otorhinolaryngol. 2019; 116: 65-9.
3. Hamouda S, Oueslati A, Belhadj I, Khalsi F, Tinsa F, Bousseta K. Flexible bronchoscopy contribution in the approach of diagnosis and treatment of children's respiratory diseases: the experience of a unique pediatric unit in Tunisia. Afri Health Sci. 2016; 16: 51-60.
4. Landry A, Rutter M. Airway anomalies. Clin Perinatol. 2018; 45: 597-607.
5. Lin Y, Lee Y, Jeng M, Chen W, Tsao P, Chan I, et al. Flexible bronchoscopic findings and the relationship to repeated extubation failure in critical children. J Chin Med Assoc. 2018; 81: 804-10.
6. Kocyildirim E, Kanani M, Roebuck D, Wallis C, McLaren C, Noctor C, et al. Long-segment tracheal stenosis: slide trache-

oplasty and a multidisciplinary approach improve outcomes and reduce costs. J Thorac Cardiovasc Surg. 2004; 128: 876-82.

7. Torre M, Carlucci M, Avanzini S, Jasonni V, Monnier P, Tarantino V, et al. Gaslini's tracheal team: preliminary experience after one year of paediatric airway reconstructive surgery. Ital J Pediatr. 2011; 37: 51.
8. Eber E, Antón-Pacheco JL, de Blic J, Doull I, Faro A, Nenna R, et al. ERS statement: interventional bronchoscopy in children. Eur Respir J. 2017; 50(6): 1700901.
9. Hoetzenecker K, Scweiger T, Schwarz S, Roesner I, Leonhard M, Denk-Linnert D-M, et al. Summarized institutional experience of paediatric surgery. Eur J Cardiothorac Surg. 2016; 49: 1119-26.
10. Maeda K. Pediatric airway surgery. Pediatr Surg Int. 2017; 33: 435-43.
11. Kamran A, Jennigs RW. Tracheomalacia and tracheobronchomalacia in pediatrics: an overview of evaluation, medical management, and surgical treatment. Front. Pediatr. 2019; 7: 512.
12. Henry B. Hsieh W, Sanna B, Vikse J, Taterra D, Tomaszewski K. Incidence, risk factors, and comorbidities of vocal cord paralysis after surgical closure of a patent ductus arteriosus: a meta-analysis. Pediatric Cardiology. 2018; 40: 116-25.
13. Liu Y. Varier I. Onkasuwan J. Use of audiometric measurement for assessment of vocal-fold function in postextubation infants. JAMA Otolarymgol Head Neck Surg. 2017; 143: 908-11.
14. Shen S, Chang H, Chang J, Hsu C, Peng C, Lee K. Bilateral vocal gold immobility in a single tertiary hospital in northern Taiwan: A 23-year retrospective review. Medicine (Baltimore). 2019; 98(9): e14691.