Endotracheal Intubation in a Down Syndrome Adult Undergoing Cataract Surgery – a Multidisciplinary Approach

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ABSTRACT

The present case report aims to describe and discuss the approach for the management of difficult endotracheal intubation in an adult with Down syndrome undergoing cataract surgery. A 26-year-old female with Down syndrome and a validated diagnosis of cataract requiring surgery was examined in order to assess the degree of difficulty of endotracheal intubation. Patients with Down syndrome have characteristic craniofacial abnormalities which require a thorough pre-operative assessment to anticipate and prepare for a difficult endotracheal intubation. Before the surgery, a series of clinical and paraclinical examinations were conducted. Although cataract surgery generally requires loco-regional anesthesia, in our case it was performed under general anesthesia. Indicators of potentially difficult intubation were macroglossia, prognathism, short neck, limited degree of head extension and obesity. The pre-operative examinations, which revealed a high degree of endotracheal intubation, allowed the anesthetist to achieve a better peri- and intra-operative management of the patient.

Keywords: endotracheal intubation, Down syndrome, lateral cephalogram

BACKGROUND

Trisomy 21, meaning an extra copy of human chromosome 21, gives rise to a characteristic physical and cognitive phenotype. In terms of medical pathology, individuals with Down syndrome have an increased risk of developing several medical conditions. Some of the diseases that occur more often are heart defects, vision problems, hearing loss, infections, hypothyroidism, blood disorders, hypotonia, sleep disorders, gum and dental problems or epilepsy (1). Therefore, they have particular medical needs and require a special care. The preliminary anesthetic examination should involve careful evaluation of these patients for achieving a safe surgery (2).
The aim of this case report is to evaluate the type of anesthesia required in an adult patient with Down syndrome (DS) undergoing cataract surgery. Furthermore, the elements necessary for assessing the degree of difficulty of endotracheal intubation were evaluated through a multidisciplinary approach.

**CASE REPORT**

A 26-year-old female, confirmed cytogenetically at birth with a full trisomy 21, presented to a private Ophthalmology Clinic in Bucharest accusing severe decrease in visual acuity. After the ophthalmological evaluation, the patient was diagnosed with cataract, which is frequently encountered disease in DS subjects (3). For this condition, the patient was assigned to undergo cataract surgery in order to improve visual acuity.

Phenotypically, the facial examination of the patient reveals the specific features in a DS subject (4): upwardly and outwardly oriented oblique fissure vents, epicanthic folds, strabismus, small nose with a flattened root, short neck, broad hands with short fingers, brachydactyly in finger V of the upper limbs and macroglossia (Figure 1).

The Ethics Committee of “Carol Davila” University approved the clinical protocol and the informed consent form for this study. The research has been conducted in full accordance with the World Medical Association Declaration of Helsinki. The written informed consent was obtained from the patient’s parents.

The pre-anesthetic examination included taking the family and medical history of the patient, which was correlated with a series of clinical and paraclinical evaluations.

The family history revealed that the patient has two healthy sisters with normal phenotypes who gave birth to clinically normal children. Between the two sisters and the patient, the parents had a healthy boy at birth, but who died from meningoencephalitis three months later. The parents’ ages (the mother 30 years old, the father 31 years old) at the birth of the child with DS are not considered etiopathogenic factors for the genesis of the syndrome. By analyzing the family tree, it appears that the patient is a sporadic case (Figure 2).

Therefore, in order to assess the difficulty of endotracheal intubation, several evaluations were carried out. The weight, height, body mass index (BMI), neck circumference, maximum mouth opening, thyromental distance (Patil test),

![FIGURE 1](image1.png)

**FIGURE 1.** Clinical phenotype showing the distinct facial and physical characteristics of a DS patient: (A, B) frontal and profile facial appearance; (C, D) frontal and profile picture showing small stature, short neck, obesity; (E) the hands are broad and flat, with short fingers; the little finger slants inward.

![FIGURE 2](image2.png)

**FIGURE 2.** Family tree of the patient with DS (II7) indicating the patient as a sporadic case. The patient has no relevant previous illnesses that could place her in a risk group for the anesthetic procedures. This condition, along with the fact that the cooperation with the patient was difficult, were the rationale for performing this surgical procedure under general anesthesia.
sternomental distance (Savva test) were recorded, and the Mallampati score was assessed.

The clinical evaluation began by measuring the patient’s height and weight in order to calculate the BMI (Table 2). So, the patient was included in the third class of obesity (5), which increased the difficulty of intubation by altering the position of the glottis and limiting the mobility of the cervical spine.

The neck circumference measured below the laryngeal protrusion and perpendicular to the long axis of the neck (6) was 43 cm. This parameter is an indicator for the limited mobility of the neck, which is a predictive element for a difficult intubation by reducing the head extension.

Furthermore, the thyromental distance was measured (Patil test) between the top of the thyroid cartilage and the Mentalis point with the head in full extension (7). The measured distance of 6 cm was another important risk factor for intubation, given the fact that values of <6.5 cm were shown to predict a difficult intubation (8, 9).

Sternomental distance (Savva test) was appreciated between the sternal notch and the Mentalis point, also with the head in full extension (7). Considering that the measured distance was 12 cm and values less than 12.5 cm increase the difficulty of endotracheal intubation (9, 10), this parameter is another indicator of a potential complicated procedure.

Intraorally, the maximum mouth opening was measured between the incisal edges of the upper and lower canines. A mouth opening under 30 mm is a predictor of difficult intubation (9). It was observed that in this case, the distance was 38 mm.

Tooth mobility was also evaluated according to Miller’s classification, which comprises four classes (11): (0) no movement detected; (1) the tooth may be moved less than 1 mm in buccolingual or mesiodistal direction; (2) the tooth may be moved 1 mm or more in buccolingual or mesiodistal direction; (3) the tooth may be moved 1 mm or more in buccolingual or mesiodistal direction, but mobility in the occluso-apical direction is also present. Dental problems involved both upper and lower arches. The upper incisors were missing and the upper left canine fitted into class 3 mobility. In the lower arch, the incisors belong to class 1 mobility.

The patient was assigned to class IV Mallampati due to the lack of visibility of the soft palate (Figure 3).

Also, the Cormack-Lehane score was determined by visualizing the appearance of the larynx through direct laryngoscopy. It designates four grades of difficulty, with the same significance to the previous score (8). The patient was classified into grade 4 of difficulty due to the lack of visibility of the epiglottis.

<table>
<thead>
<tr>
<th>Anthropometric point</th>
<th>Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mentalis (Me)</td>
<td>The most inferior point of the mandibular symphysis</td>
</tr>
<tr>
<td>Condilion (Co)</td>
<td>The point built at the intersection between the tangent to the posterior side of the condyle and the mandibular angle with the tangent to the most upper point of the condyle</td>
</tr>
<tr>
<td>Gnatio (Gn)</td>
<td>The lowest and most posterior point of the mandibular symphysis</td>
</tr>
<tr>
<td>Anterior nasal spine (ANS)</td>
<td>The top of the anterior nasal spine</td>
</tr>
<tr>
<td>Posterior nasal spine (PNS)</td>
<td>The top of the posterior nasal spine</td>
</tr>
</tbody>
</table>

**TABLE 1.** Anthropometric points used for clinical examination and for tracing the lateral skull radiograph

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Reference values</th>
<th>Measured value</th>
<th>Unit</th>
</tr>
</thead>
<tbody>
<tr>
<td>Weight</td>
<td>-</td>
<td>92</td>
<td>Kg</td>
</tr>
<tr>
<td>Height</td>
<td>-</td>
<td>147</td>
<td>m</td>
</tr>
<tr>
<td>BMI (5)</td>
<td>20 – 24.9</td>
<td>46.2</td>
<td>Kg/m²</td>
</tr>
<tr>
<td>Neck circumference (13)</td>
<td>&lt;43</td>
<td>43</td>
<td>cm</td>
</tr>
<tr>
<td>Thyromental distance (8, 9)</td>
<td>&gt;6</td>
<td>6</td>
<td>cm</td>
</tr>
<tr>
<td>Sternomental distance (9,10)</td>
<td>&gt;12.5</td>
<td>12</td>
<td>cm</td>
</tr>
<tr>
<td>Maximum mouth opening (9)</td>
<td>&gt;30</td>
<td>38</td>
<td>mm</td>
</tr>
<tr>
<td>Dental mobility (11)</td>
<td>0</td>
<td>3</td>
<td>class</td>
</tr>
<tr>
<td>Mallampati score</td>
<td>-</td>
<td>IV</td>
<td>class</td>
</tr>
<tr>
<td>Cormack Lehane</td>
<td>-</td>
<td>4</td>
<td>grade</td>
</tr>
</tbody>
</table>

**TABLE 2.** Parameters measured during the clinical examination and the results obtained for the DS patient
The results obtained for the parameters evaluated during the clinical examination are summarized in Table 2.

Moreover, a series of parameters on the lateral cephalogram radiograph were evaluated, whilst maintaining the subject’s head in the natural head position. The radiographs were traced on acetate paper, using conventional antropometric landmarks and measurements were carried out with a Cephalometric Protractor from 3M Unitek (Figure 4, Table 1).

![Figure 4. Cephalometric analysis showing increased mandibular effective length, normal values for C0-C1 and C1-C2 gaps and the low maxillo-pharyngeal angle.](image)

The mandibular effective length from McNamara analysis was measured between Condylion and Gnathion (Co-Gn). Normal average values are $120.2 \pm 5.3$ mm for females and $134 \pm 6.8$ mm for males (14). For the referred patient, the measured distance was 190 mm, which denoted an increased mandible and a potential difficult intubation (8).

Furthermore, the atlanto-occipital gap between C0-C1 vertebrae was appreciated by drawing a perpendicular line from the base of the occipital bone to the most posterior and superior point of the first cervical vertebra. For this parameter, normal values are between 4-9 mm (15). C0-C1 gap is a major factor that influences the extension of head because the greater the distance, the more space is available for the mobility of the head, which ensures a favorable axis for laryngoscopy and endotracheal intubation (8). In this case, the value obtained for C0-C1 gap was 9 mm, meaning that from this point of view there was a favorable circumstance for intubation.

C1-C2 gap (atlas-axis) represents the perpendicular distance between the lowest and most posterior point of the posterior arch of atlas and the most superior and posterior point of the spinous process of axis (15). The measured distance was 6 mm, representing an average value for this parameter.

Maxillo-pharyngeal angle (Delegue) is the angle between maxillary axis (MA) and pharyngeal axis (PA). MA (16) is the line passing between anterior and posterior nasal spine points (ANS, PNS), while PA is the line passing through the anterior part of the first and second cervical vertebrae (10). The value obtained for this parameter was 92°, which was predictive for difficult intubation, an angle smaller than 90° being associated with an impossible laryngoscopy.

The results obtained from the lateral cephalogram tracing are shown in Table 3.

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Landmarks</th>
<th>Normal value</th>
<th>Measured value</th>
<th>Unit</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mandibular effective length</td>
<td>Kdl–Gn</td>
<td>120</td>
<td>190</td>
<td>mm</td>
</tr>
<tr>
<td>Atlanto-occipital gap</td>
<td>C0-C1</td>
<td>4-9</td>
<td>9</td>
<td>mm</td>
</tr>
<tr>
<td>Atlas-axis gap</td>
<td>C1-C2</td>
<td>4-9</td>
<td>6</td>
<td>mm</td>
</tr>
<tr>
<td>Maxillo-pharyngeal angle</td>
<td>MA–PA</td>
<td>100</td>
<td>92</td>
<td>degree</td>
</tr>
</tbody>
</table>

Following the clinical and paraclinical examinations performed, it can be assumed that for this patient, endotracheal intubation presented a high degree of difficulty. For this reason, we chose the video laryngoscope (C-MAC, Macintosh laryngoscope blade, size 3) to achieve the intubation.

During induction of general anesthesia, the patient received Fentanyl 2 microg/kg-body weight, Propofol 2 mg/kg-body weight, Succinylcholine 1 mg/kg-body weight and adequate mask ventilation. The endotracheal tube, size 7.0, was advanced without difficulty after identifying the anatomic landmarks (the tongue, the uvula, the epiglottis, the arytenoid cartilages and the vocal cords). Capnography and auscultation confirmed the correct placement of the tube into the trachea.

In this case, intubation was performed with video laryngoscopy, which allowed us to achieve a detailed examination of the larynx in order to attain a better management in a difficult airway situation.
DISCUSSIONS

For a cataract surgery, the protocol usually involves a loco-regional anesthesia that is retrobulbar or peribulbar (17). Contraindications to use this technique are related to allergies to anesthetic agents and the presence of ocular infections. Although loco-regional anesthesia would have been the choice in another similar surgical procedure, the intubation was performed under general anesthesia due to the patient’s low compliance.

The evaluation carried out in order to assess the difficulty of endotracheal intubation consisted of eight clinical parameters and four paraclinical elements. The results showed that seven clinical and two paraclinical parameters could be incriminated as high risk factors for the endotracheal intubation. This allowed us to conclude that there was the investigated patient fell into the category of high degree difficulty at intubation.

The lateral cephalogram radiograph is not routinely used to assess the upper airway, but it could be a valuable tool in determining the difficulty of endotracheal intubation taking into consideration that no single test can provide a high index of sensitivity and specificity (10).

In the literature, there are several studies that describe other parameters in order to assess the difficulty of endotracheal intubation in a preoperative evaluation of a patient with Down syndrome. One of these studies uses the magnetic resonance imaging to determine the proper size of the endotracheal tube in DS subjects due to an overall decrease in the diameter of the tracheal lumens (18). In addition, the authors opted for an endotracheal tube at least two sizes smaller than the one used in a healthy child (18).

Other authors recommend that the intubation probe could be selected by referring its dimension to the size of the earbuds or to the patient age (19), but this cannot be applied in subjects with DS because they have brachydactyly or clinodactyly in finger V and present a slower growth rate (hypotrophy).

A difficult intubation was reported in a patient with DS and lingual tonsillar hypertrophy. The alternative in this case was to intubate the patient with a nasotracheal tube by fibroscopy (20).

CONCLUSIONS

From this study, it was concluded that patients with trisomy 21 are particular cases in medical surgical pathology, due to the anatomical and physiological changes. Down syndrome is a common finding in human pathology, both in adults and in children, and it requires a multidisciplinary medical evaluation. Special care must be taken because each person with DS is affected differently and has unique health issues and variability in the severity of symptoms.

Given the fact that studying the degree of endotracheal intubation difficulty in subjects with Down syndrome is a topic less debated in literature, more studies are needed to assess the anesthetic particularities of these patients.

Conflicts of interest: I undersign, certificate that I do not have any financial or personal relationships that might bias the content of this work.

Statement of Human Rights: I undersign, certificate that the procedures and the experiments I’ve done respect the ethical standards in the Helsinki Declaration of 1975, as revised in 2000 (5), as well as the national law.

Informed consent statement: I undersign, certificate that I have the written consent of the patient’s legal guardian in order to present the case in this scientific paper.

REFERENCES


