Anesthetic Management of a Down Syndrome Patient with Subocclusive Syndrome

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ABSTRACT

The present study aims to describe the anesthetic management of a patient with Down syndrome in connection to the degree of difficulty in orotracheal intubation and the associated risks of general anesthesia. The established diagnosis was subocclusive syndrome, requiring an emergent surgical intervention. Preoperatively, the patient was stabilized, and secondly, a series of clinical and paraclinical investigations were carried out in order to assess the best management of the anesthetic procedure. Risk factors were identified, including ischemic heart disease, grade II obesity, hyperglycemia, patient’s old age and marked mental retardation. The results of examinations that were carried out showed that the degree of difficulty of orotracheal intubation was low. Finally, the preoperative assessment indicated that the most appropriate approach for this patient was to perform a general anesthesia with endotracheal intubation. However, special care was given to the anesthetic and postoperative condition of the patient in order to lower the possible complications of the surgical intervention.

Keywords: emergency, surgery, Down syndrome, orotracheal intubation.

INTRODUCTION

Down syndrome (DS), a well-known chromosome disorder, affects millions of people worldwide. Average life expectancy of people with DS has increased in the last decades (1, 2) due to improved medical care. An adult with DS can reach an old age such as 50s, 60s or more (3, 4). But old age can bring unexpected medical problems for both DS people and health care providers. Being one of the most common genetic disorders, every clinician of any specialty can expect to deal with some medical problems (vision and hearing impairment, thyroid dysfunction, sleep apnea, celiac disease, osteoarthritis, osteoporosis, atlantoaxial instability or Alzheimer’s disease) related to DS. Some of these medical problems require emergency care and the medical team involved needs to assess DS cases with respect to their general and particular conditions.
Therefore, the aim of this case report is to assess the management of an adult patient with Down syndrome by the anesthetic and surgical team in order to diagnose and treat a subocclusive syndrome. Yet, special attention was given to analyzing the degree of difficulty of the endotracheal intubation and increased risks for complications associated with general anesthesia.

The Ethics Committee of “Carol Davila” University of Medicine and Pharmacy, Bucharest, Romania, approved the clinical protocol and 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 aunt, who was his legal guardian.

**CASE REPORT**

A 51-year-old male patient has presented to the emergency room of a public hospital in Bucharest for general health alteration, fever (38.5°C), absence of intestinal transit and episodes of vomiting, which started five days before presentation. In order to form a diagnosis of admission, the patient was investigated clinically and paraclinically.

The first element that appealed our attention was the phenotype aspect of an individual with DS. The face was small, with a round shape and a flattened profile due to maxillary hypoplasia. Also, the cephalic extremity showed occipital flattening and strabismus (5) was easily observed. The specific epicanthus was present while the eyebrows were oblique, upward and outwardly oriented. The nose was small, with a flattened root. The ears were also small and inferiorly positioned in respect to the external angle of the eyes. Microstomia with dropped commissures were present, as well, and the tongue was protruding, with a scrotal aspect. In the oral cavity, microdontia and anodontia were noticed. The neck was short with an excess of adipose tissue. The upper and lower extremities were small, wide and flat (3, 6). Also, the patient had a marked mental retardation, which is frequently encountered in DS patients after the age of 30 because it is associated with Alzheimer disease (7-9). All these characteristics were used to diagnose trisomy 21, which was confirmed by the cytogenetic test.

The patient’s family medical history indicated that this individual was a sporadic case of trisomy 21. From the heredo-collateral antecedents, it could be mentioned that the patient’s parents and his only brother had a history of ischemic heart disease associated with dyslipidemia and diabetes, and they had both died from heart attack at early ages.

The personal pathological antecedents included hypertension, ischemic heart disease and grade II obesity, with the following background medication: trimetazidine, ranitidine and metoclopramide. Although DS individuals frequently present malformations of the heart and gastrointestinal tract (10), this patient had no surgical history, which complicated the anesthetic and surgical approach due to the lack of elements that could anticipate potential risks and a difficult intubation.

The clinical examination revealed a conscious patient, with psychomotor agitation, hemodynamically and respiratory stable (blood pressure 120/70 mmHg, tachycardic with 107 heart rate, \(\text{SpO}_2 97\%\)). Instead, a tumoral formation of about 25/20 cm could be palpated at the abdominal level, which was painful, irreducible, but with no signs of peritoneal irritation. It should be taken into consideration that in DS patients, acute pain perception is delayed, which makes the pathological process to be discovered in a late phase of evolution and further complications can be expected (11).

Paraclinical examinations showed hyperbilirubinemia (total bilirubin 1.4 mg/dL), hyperglycemia (132 mg/dL), diselectrolyemia based on hyponatremia (128.3 mmol/L) and hypokalemia (3.5 mmol/L). Ultrasound examination showed meteorism, without significant changes in the hepatobiliary system, spleen, pancreas, kidneys and costo-diaphragmatic sinuses. A further CT examination revealed a parietal defect on the median line, with a 60 mm cross-sectional diameter, which was highly suggestive for a strangulated umbilical hernia.

Treatment plan aimed to repair the parietal defect by herniorrhaphy. The preanesthetic assessment included a series of clinical and paraclinical investigations in order to determine the degree of difficulty in endotracheal intubation and to prepare the patient for surgery (12).

The patient’s body weight was 110 kg and height 170 cm, corresponding to a BMI of
38 kg/m² and grade II obesity, which are important risk factors for difficult intubation. The neck circumference was 50 cm and the abdominal circumference 134.5 cm, which also suggested a difficult intubation. The sternomental distance (Sava test) was 15 cm and the thyromental distance (Patil test) 6.5 cm (Table 1). These two tests together with a maximum mouth opening of about 7.5 cm and the Mallampati II score were arguments for the absence of a difficult intubation.

Because the parameters needed to evaluate the difficulty of endotracheal intubation were not homogenous, a series of elements were prepared in order to assist the orotracheal intubation before surgery: laryngoscope with various blades, probes of various sizes, flexible catheter, endotracheal tubes, laryngeal mask, tracheostomy kit and fibroscope. The larynx mask was not used by the first intention due to the increased risk of bronchial-pulmonary aspiration occurring in a patient with such a conformation (abdominal circumference 134.5 cm), but also because of the surgical pathology (13).

Preoperatively, the patient was conscious, relatively cooperative, hemodynamically and respiratory stable (102/60 mmHg blood pressure, 75 heart rate, SpO₂ 97%). At this time, the patient was fitted a 18G-sized peripheral venous catheter (PVC), enabling to administer approximately 500 mL of 0.9% physiological saline, 3 mg of midazolam and 1.5 g of cefuroxime. A nasogastric probe was also mounted.

In the operating room, the patient was pre-oxygenated for five minutes with 100% oxygen with a flow of 6 L/min. This led to an increase in the oxygen reserve in the pulmonary system and SpO₂ levels from 97% to 98%.

The anesthetic induction in the rapid sequence consisted of fentanyl 150 µg and propofol 150 mg. When the contact with the patient was lost, the Sellick maneuver was used in order to reduce the risk of bronchopulmonary aspiration.

A depolarising agent – suxamethonium chloride (100 mg) – was administered to facilitate orotracheal intubation. It should be noted that we did not have any contraindications for the succinylcholine, which often occurs in DS patients because of high values of intraocular pressure (14).

During direct laringoscopy with size 4 Macintosh blade, the posterior laryngeal aperture was visualized which corresponds to a grade 2 Cormack-Lehane score. The orotracheal intubation was performed without incidents and it was validated by pulmonary auscultation and capnography.

A size of 7.5 was chosen for the intubation probe (Figure 1), being known that people with

![FIGURE 1. Intraoperative aspects of the surgical procedure: the patient is intubated with a 7.5-sized probe during herniorrhaphy](image-url)
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Trisomy 21 have stenosis at the tracheal level (15) and for this reason it is advisable to use probes with a smaller diameter. Some clinical trials have found a correlation between the size of the intubation probe and that of finger V in hand, but in this case we have not been able to use this element because finger V of the hand was much smaller in size and had an atypical form (16).

The intraoperative monitoring was standard by non-invasive measuring of the blood pressure, the ECG, the heartbeat, the temperature and the pulse oximetry. Anesthesia was maintained with fentanyl 200 µg, atracurium 40 mg and sevoflurane 2%, with a flow of 4 L/min 50:50 oxygen:air. Shortly after induction, blood pressure decreased from 115/70 mmHg to 85/50 mmHg, so 500 mL of colloid were administered.

Intraoperatively, no incidences have occurred. The blood pressure was maintained around 100/70 mmHg, with a heart rate of 70. The duration of the surgery was 1 hour and 40 minutes (Figure 2) and an arterial blood gas (ABG) sample collection was obtained one hour after surgery, which showed that hiponatremia had persisted (Table 2).

Postoperatively, the patient was conscious, relatively cooperative, apyretic, hemodynamically stable, and the ABG values are shown in Table 2.

<table>
<thead>
<tr>
<th>Medication</th>
<th>Dose</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ringer</td>
<td>1000 mL</td>
</tr>
<tr>
<td>Glucose 10% + 10 IU</td>
<td>1000 mL</td>
</tr>
<tr>
<td>Insulin/500 mL</td>
<td></td>
</tr>
<tr>
<td>Gastric protector</td>
<td>1f/12 h</td>
</tr>
<tr>
<td>Prokinetic</td>
<td>1f/12 h</td>
</tr>
<tr>
<td>Anticoagulant</td>
<td>0,4 mL/sc/24 h</td>
</tr>
<tr>
<td>Antibiotic</td>
<td>1,5 g</td>
</tr>
<tr>
<td>Analgesic</td>
<td>1f/8 h</td>
</tr>
<tr>
<td>Antipyretic</td>
<td>1f/12 h</td>
</tr>
<tr>
<td>NSAIDs</td>
<td>1f/12 h</td>
</tr>
</tbody>
</table>

**Table 2. Postoperative treatment**

**Figure 2.** Down patient after surgery (A) and in the postoperative care room (B)

### Table 2. ABG values one hour after surgery

<table>
<thead>
<tr>
<th>ABG</th>
<th>Values</th>
</tr>
</thead>
<tbody>
<tr>
<td>pH</td>
<td>7,31</td>
</tr>
<tr>
<td>pCO₂</td>
<td>44 mmHg</td>
</tr>
<tr>
<td>pO₂</td>
<td>78 mmHg</td>
</tr>
<tr>
<td>Na</td>
<td>125 mEg/L</td>
</tr>
<tr>
<td>K</td>
<td>3,5 mEg/L</td>
</tr>
<tr>
<td>Ca</td>
<td>0,97 Mmol/L</td>
</tr>
<tr>
<td>Glu</td>
<td>105 Mg/dL</td>
</tr>
<tr>
<td>Lac</td>
<td>0,9 Mmol/L</td>
</tr>
<tr>
<td>Hct</td>
<td>42%</td>
</tr>
<tr>
<td>HCO₃</td>
<td>21,6 Mmol/L</td>
</tr>
<tr>
<td>BE</td>
<td>-4,1 Mmol/L</td>
</tr>
<tr>
<td>SO₂</td>
<td>94%</td>
</tr>
<tr>
<td>HBc</td>
<td>13 g/dL</td>
</tr>
</tbody>
</table>

**Table 2.** ABG values one hour after surgery
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M. B. C. and Respiratory Stable. Treatment for the 24 hours following surgery is shown in Table 3. Early oral nutrition occurred after 24 hours postoperatively and the intestinal transit after approximately 48 hours.

**DISCUSSION**

The protocol for preoperative preparation took into account that the patient had a heart condition associated with a hydro-electrolyte disorder. Considering that the patient had glycemic values above the maximum accepted limit and in his family there was a history of diabetes, we may discuss a possible type 2 diabetes. In the literature, incidence of type 2 diabetes in DS patients is lower than in the general population, but type 1 diabetes is more common (19). However, in this case, the elevated glucose levels can be related to the patient’s obesity, which increases insulin resistance.

In terms of the surgical procedure, herniorrhaphy is usually performed under rachianesthesia (20), especially when a difficult intubation is anticipated. But taking into consideration the low degree of cooperation (due to the underlying condition), psychomotor agitation, emergency situation, possible complications that may arise during surgery (increasing the surgical duration or extending the wound in the supraumbilical area, etc.), and a possible atypical anatomy occurring in patients with trisomy 21 (nostrils with small diameter, microstomia, macroglossia, dento-maxillary anomalies, cervical spine disorders, increased neck circumference, increased abdominal circumference, anatomical changes in the airways, etc.), it was decided that a balanced anesthesia using inhalation agents should be considered. Combined anesthesia (general and loco-regional anesthesia) can also be performed because it can be used as a method of postoperative analgesia too.

In the literature, emergency care for patients with DS is scarcely discussed. Due to the high prevalence of congenital heart disease, emergency treatment should include airway management, supplemental oxygen and IV access (17, 18). Also, in patients with heart failure, it is recommended to carefully administer diuretics and balance the blood volume (17, 18). In our case, the emergency situation was dealt by stabilizing the hydro-electrolyte values in order to lower the risks that can be associated with the surgical procedure.

**CONCLUSION**

Anesthetic management in patients with DS requires special consideration due to the anatomical alterations and particular pathology encountered. For the case considered in this study, general anesthesia with orotracheal intubation was selected for the surgery procedure. The parameters analyzed in order to assess the degree of difficulty in endotracheal intubation have suggested that this manoeuvre was scarcely influenced by changes in the cephalic extremity. But special care was necessary because the patient presented a cardiac disease associated with dyslipidemia and hyperglycemia, which could increase the risk of a heart attack. The patient’s age associated with a severe degree of mental retardation represented key elements in choosing the type of anesthesia.

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Human Rights Statement: All procedures and experiments met the ethical standards in the Helsinki Declaration of 1975, as revised in 2000 (5), as well as the national law.
Patient Informed Consent: The written consent of the patient’s legal guardian was obtained.
REFERENCES


