Chapter 15
Down Syndrome (Pediatric)

Claire Sampankanpanich Soria

Case Outline

Learning Objectives

1. Discuss risks of anesthesia in patients with Down syndrome.
2. Discuss pre-operative evaluation of patients with Down syndrome.
3. Discuss intraoperative precautions to take with Down syndrome.

Simulator Environment

1. Location: pre-operative area of an adult hospital
2. Manikin setup:
   (a) Age: adult
   (b) Lines: none at start of case
   (c) Monitors: none on patient at start of case
3. Medications available: normal saline, propofol, etomidate, succinylcholine, rocuronium, epinephrine, albuterol, fentanyl, midazolam, ketamine.
4. Equipment available

C. Sampankanpanich Soria (✉)
University of California, San Diego Medical Center, San Diego, CA, USA
e-mail: cssoria@health.ucsd.edu

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(a) Airway equipment: ventilator, face mask, laryngoscope and cuffed and uncuffed endotracheatl tubes (ETTs) of various sizes, stylets, oral airway, nasal trumpet, laryngeal mask airway (LMA), suction.
(b) Monitors: pulse oximeter, blood pressure cuff, 5-lead electrocardiogram (EKG).
(c) Lines: arterial line kit, central line kit, peripheral intravenous (PIV) kits
(d) Crash cart with defibrillator
(e) Paperwork: pre-operative anesthesia history and physical

Actors

1. Scrub tech
   (a) The scrub tech is busy opening trays and making lots of noise.
2. Circulator nurse
   (a) The nurse is busy helping to open trays.
3. Surgeon
   (a) The surgeon is busy dictating the operating report from the previous patient.

Scenario Development

1. Background
   (a) You are the anesthesiologist starting the case of a 26-year-old boy with Down syndrome, severe developmental delay, history of large ventricular septal defect (VSD) status post repair in infancy, obesity, and obstructive sleep apnea (OSA) noncompliant with continuous positive airway pressure (CPAP) due to discomfort with face mask. He is scheduled to go to the OR for tonsillectomy and adenoidectomy.
2. Phase 1: patient refusal to leave the preoperative area, parent induction
   (a) In the preoperative area, the patient is calm and quiet, sitting comfortably in the gurney with warm blankets, with mom and dad at bedside. He had refused to allow the nurses to check his vital signs or to place a PIV.

(b) The parents ask the anesthesiologist if there is any way they can do the PIV asleep, since he is scared of needles and has been a difficult PIV placement in the past.
(c) The learner may offer to do a mask induction and place the PIV asleep, and may consider calling for difficult PIV equipment to be sent to the room ahead of time (e.g. ultrasound, vein finder).
(d) When it is time to leave for the operating room (OR), the patient refuses, starts crying, and refuses to let go of his mother.
(e) The parents ask the anesthesiologist if it is possible to do a parent induction. They have always done parent inductions in the past. It is their son's first time receiving surgery in an adult hospital.
(f) The learner may offer to do a parent induction. If they choose to do a parent induction, they should provide clear instructions for what to expect during a mask induction and make sure the parent will be calm and willing to leave when asked to do so.
(g) With a parent present, the patient will be calm and cooperative.

3. Phase 2: upper airway obstruction and bradycardia during induction

(a) During induction, the patient will develop significant upper airway obstruction making mask ventilating challenging.
(b) The parent will become anxious and ask what's wrong. The learner should provide reassurance to the parent that this is expected and under control. The learner should ask the parent to leave so they can focus on patient care.
(c) The learner may cautiously place airway adjuncts such as an oral airway or nasal trumpet to assist with mask ventilation, but recognize that the patient may be in stage 2 and develop laryngospasm.
(d) The learner may administer CPAP by closing the airway pressure release valve and holding pressure using the ventilator bag.
(e) The learner should try to establish IV access as soon as possible but avoid triggering laryngospasm during stage 2.
(f) The PIV placement will be challenging and require multiple attempts. During attempted PIV placement, the patient will become bradycardic.
(g) The learner should recognize the bradycardia and discontinue the nitrous oxide, decrease the sevoflurane, and take over ventilating the patient manually via bag valve mask with 100% fraction of inspired oxygen (FiO_2).
(h) A PIV will be placed and the learner will proceed with intubation uneventfully.

Scoring Rubric

Table 15.1 Scoring rubric for case scenario on Down Syndrome

Topic: Down Syndrome			
Participant Name:			
Evaluator Name:			
Score:			
		Completed	Not Completed
Pre-operative area			
Communication	Asks parents about history of previous anesthetics/inductions and what's worked well for patient in past		
	Counsels parents about pros/cons and sequence of events of parent induction		
	Prepares room with equipment for difficult peripheral intravenous (PIV) line placement (ultrasound, vein finder)		
Upper airway obstruction			
Evaluation and management	Creates calm, quiet, attentive environment during induction		
	Identifies severe upper airway obstruction		
	Double checks / repositions hands to optimize mask seal		
	Provides continuous positive airway pressure (CPAP) via circuit/mask		
	May cautiously place oral airway and/or nasal trumpet		
	Politely but firmly asks parent to leave so they can focus on patient care		
Difficult PIV and Bradycardia			
	Identifies bradycardia		
	Adjusts anesthetic: Switches to 100% fraction of inspired oxygen (FiO_2), decreases sevoflurane concentration		
	Takes over mask ventilation instead of allowing spontaneous ventilation		

Summary of Clinical Teaching Points

What are the chances that you will take care of a patient with Down Syndrome? [1, 2]

- Down Syndrome is the most prevalent genetic disorder worldwide: 1 in 800 live births; >350,000 people in the United States.
- It is associated with multiple congenital anomalies that can affect every major organ system.
- Risk factors: maternal age >35 years old; exposure to pesticides and electromagnetic fields; smoking; drinking alcohol; thyroid autoimmunity; exposure to radiation and anesthetic agents

What are the potential challenges to airway management in Down Syndrome patients? [1, 2]

Table 15.2 Potential challenges to airway management in Down Syndrome patients by upper and lower airway anatomy

Upper Airway	Lower Airway
Choanal atresia	Airway malacia
Microdontia	Short neck
Mandibular hypoplasia	Hypotonia
Adenotonsillar hypertrophy	Subglottic stenosis
Macroglossia, relative glossoptosis	Respiratory tract infections
Microcephaly	Small upper and lower airways
Sleep apnea	
Upper airway obstruction	
Obesity	
Midface hypoplasia	
Increased secretions	
Small nasopharynx	

Table 15.3 Potential airway complications and management techniques in Down Syndrome patients by intra-operative and post-operative periods

	Complications	Management
Intra-operative	Difficult intubation Difficult mask ventilation Bronchospasm Tracheal stenosis: Difficult to pass endotracheal tube	Prepare additional rescue airway equipment (video laryngoscope, flexible fiberoptic bronchoscope, intubating laryngeal mask airway, flexible Bougie, emergency surgical airway equipment) Consider downsizing endotracheal tube Ensure adequate cuff leak Awake extubation
Post-operative	Post-extubation respiratory distress, stridor, croup, bronchiolitis, cyanotic events, wheezing Airway obstruction	Longer post-anesthesia care unit stay or overnight admission

What are cardiac considerations when caring for Down Syndrome patients? [1, 2]

40–50% of patients with Down Syndrome have some type of congenital heart disease. The following table provides a cursory overview of common heart defects.

Table 15.4 Cardiac anesthetic considerations for Down Syndrome patients

System	Pathophysiology	Anesthetic Considerations
Cardiac – Acyanotic defects	Complete atrioventricular defect Ventricular septal defect Atrial septal defect	Avoid increases in pulmonary vascular resistance Avoid high systemic vascular resistance Use low fraction of inspired oxygen Avoid low end-tidal carbon dioxide Antibiotic prophylaxis
Cardiac – Cyanotic defects	Tetralogy of Fallot	Normal to increased fraction of inspired oxygen Maintain normal pH Keep end-tidal carbon dioxide normal to low Keep systemic vascular resistance within normal limits Early treatment of intraoperative "Tet spell:" fluid bolus starting 10–20 cc/kg, phenylephrine 1 mcg/kg, oxygen
Rhythm anomalies	Conduction disturbances after repair of atrio-ventricular canal defect, tetralogy of Fallot, ventricular septal defect, transposition of the great arteries	Detect bradycardia quickly Increase fraction of inspired oxygen Caution with volatile anesthetic
Pulmonary vasculature	Pulmonary hypertension Eisenmenger syndrome (associated with obstructive sleep apnea and pulmonary hypoplasia)	Avoid exacerbating pulmonary hypertension (avoid hypoxemia, hypercarbia, and acidosis)

What are the indications for spontaneous bacterial endocarditis (SBE) prophylaxis? [1, 2]

- Dental surgeries or other "dirty" surgeries (not for all routine Urology cases).
- Prosthetic cardiac valve or prosthetic material used for cardiac valve repair.
- Prior infective endocarditis.
- Congenital heart disease:
  - Unrepaired cyanotic congenital heart disease, including palliative shunts and conduits.
  - Completely repaired congenital heart disease with prosthetic material or device, whether by surgery or catheter intervention, that were repaired within the last 6 months.
  - Repaired congenital heart disease with residual defects at the site or adjacent to the site of prosthetic patch or prosthetic device (which inhibit endothelialization).
- Cardiac transplant recipients who develop cardiac valvulopathy.

What are neurologic and musculoskeletal considerations for patients with Down Syndrome? [1, 2]

- Atlanto-occipital instability
  - 20% incidence
  - Ligamentous laxity of atlanto-axial joint → C1-C2 subluxation → spinal cord injury
- Minimize degree of neck flexion, extension, and rotation, especially during intubation
- Careful intraoperative positioning
- Review neck radiographs pre-operatively, where indicated
  - Not routinely done
  - Symptoms that should raise concern for possible cervical spine instability and prompt imaging pre-operatively: neck pain, gait disturbances, hand dysfunction, dizziness, bowel/bladder dysfunction, weakness, paresthesias, pain with neck flexion and extension, hyperactive distal tendon reflexes, ankle clonus, muscle weakness, increased muscle tone, neck discomfort, abnormal gait, difficulty walking, excessive laxity of other joints (fingers, elbows, knees)
- Consider using cervical collar intraoperatively if there is demonstrated cervical spine instability

What are gastrointestinal considerations for patients with Down Syndrome? [1, 2]

Table 15.5 Other anesthetic considerations for patients with Down Syndrome by organ system

System	Feature
Neurologic	Significant hearing loss and eye and vision problems Generalized hypotonia and joint laxity Cognitively challenged children can be harder to manage during induction – Consider parental presence
Immune	Some degree of immune dysfunction → affects cellular and humoral immunity → increased rate of infection Chronic sinus and ear infections
Endocrine	Thyroid hypofunction Diabetes mellitus
Hematologic	Polycythemia – May need phlebotomy to relieve circulatory failure
Gastrointestinal Renal	Umbilical hernia Renal malformations Undescended testes, hypospadias
Vascular	Radial artery abnormalities Makes percutaneous cannulation challenging

- High risk of gastroesophageal reflux disease (GERD)
  - Vomiting
  - Esophagitis (chest pain, anemia, irritability)
  - Respiratory: apnea, coughing, wheezing, aspiration pneumonia
- Common associated congenital defects
  - Duodenal atresia
  - Tracheo-esophageal fistula
  - Hirschsprung disease
  - Imperforate anus
- Consider rapid sequence induction where indicated

What are other systemic considerations for patients with Down Syndrome? [1, 2]

References

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