

Anesthetic Management of a Patient with Trisomy 18 and Tracheoesophageal Fistula

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Objectives

- Briefly describe Trisomy 18 and the anesthetic implications for a one day old
- Review tracheoesophageal fistula
- Explore the management of a one day old patient undergoing repair of tracheoesophageal fistula

Trisomy 18 (Edwards Syndrome)

- Error in cell division → chromosomal disorder
- Characterized by a broad spectrum of variable dysmorphic features and organ malformations
- Occurs in approximately 1 out of every 2,500 pregnancies
- Extremely poor prognosis

Common Associated Problems

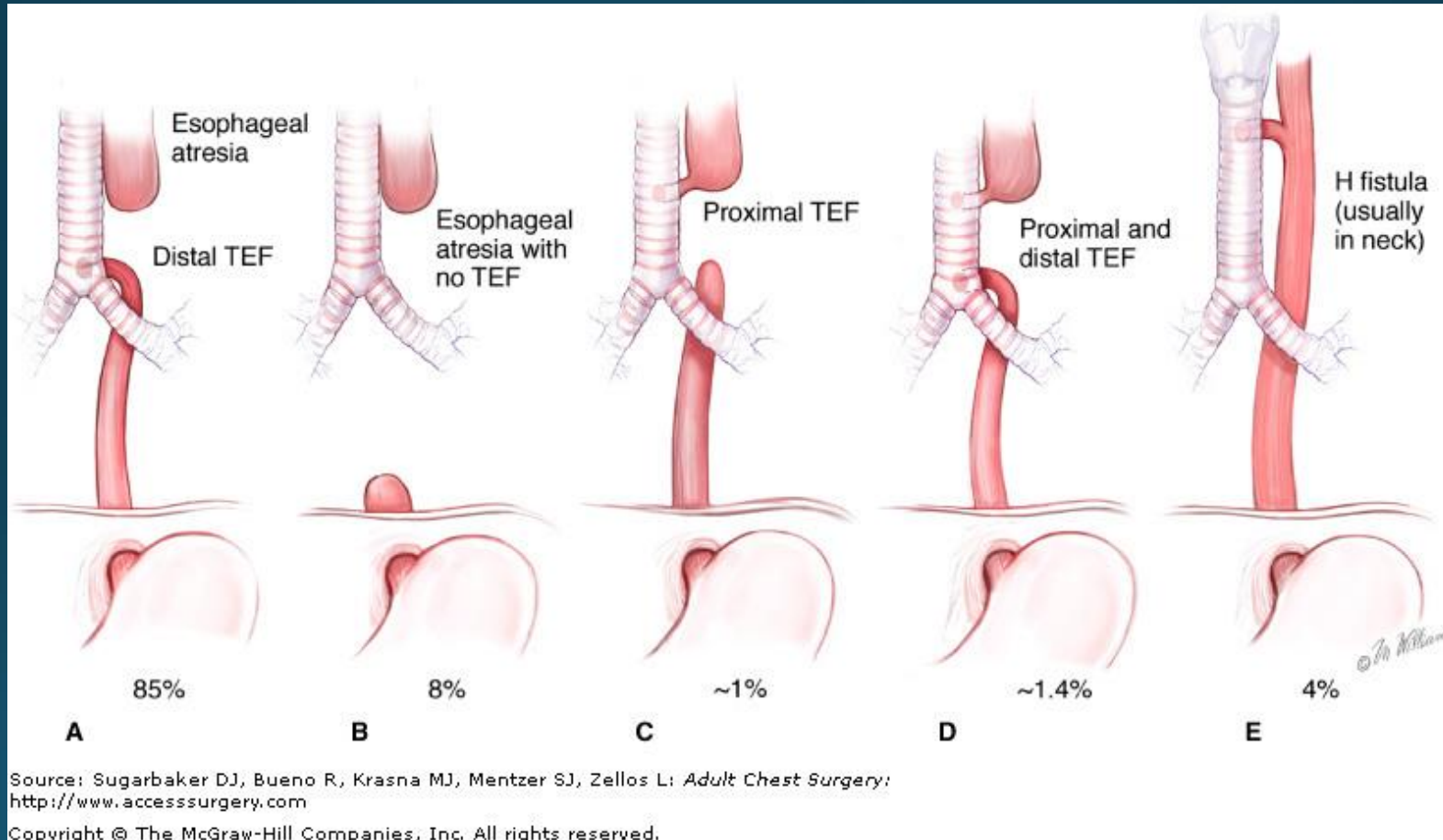
- Heart defects
 - VSD
 - ASD
 - Coarctation of the aorta
- Kidney problems
- Omphalocele
- Esophageal atresia (25%)
- Microcephaly
- Micrognathia
- Strawberry-shaped head
- Rocker bottom feet
- Polyhydramnios
- Clenched hands
- Low-set ears
- Severe developmental delays
- Umbilical or inguinal hernia



Newborn – 1 day old

- Shunts
 - Foramen Ovale
 - Ductus Arteriosus
 - Ductus Venosus
- Thermoregulation - non-shivering thermogenesis
- Airway differences
- O₂ consumption (7-8 ml/kg)
- Immature respiratory system

Tracheoesophageal Fistula



Tracheoesophageal Fistula

- Normally diagnosed after birth when an orogastric tube cannot pass into the stomach, coughing/choking after first feeding
- VACTERL syndrome
- Preoperative echo
- Standard monitors +/- invasive
- Preductal and postductal oximeter probes
- Bronchoscopy prior to intubation

Anesthetic Considerations

- Copious secretions – frequent suctioning
- Avoid PPV – maintain SV or use small Vt
- IV or inhalational induction if stable
- ETT should be above carina and below fistula to prevent gastric distention
- No neck extension during or after repair

Baby Patient

- 1 day old female, gestational age 37 weeks
- 1.5 kg
- c/s for pre-eclampsia and fetal distress (APGAR 3/8)
- Initial resuscitation: CPAP followed by SV RA
- Microcephalic, enlarged occiput, high arched palate, overlapping digits on hands, clenched fists, rocker bottom feet, generalized hypotonia, systolic murmur
- Initial plan was for palliative care

Initial Echo

- All valves thickened consistent with T18
- Large atrioventricular canal **VSD**
- Additional moderate posterior muscular **VSD**
- Redundant tricuspid valve tissue
- Non-obstructive RV muscle bundle
- Bicuspid aortic valve
- **PFO** with moderate L → R shunt
- **Large PDA with predominant L → R shunt, restrictive flow**

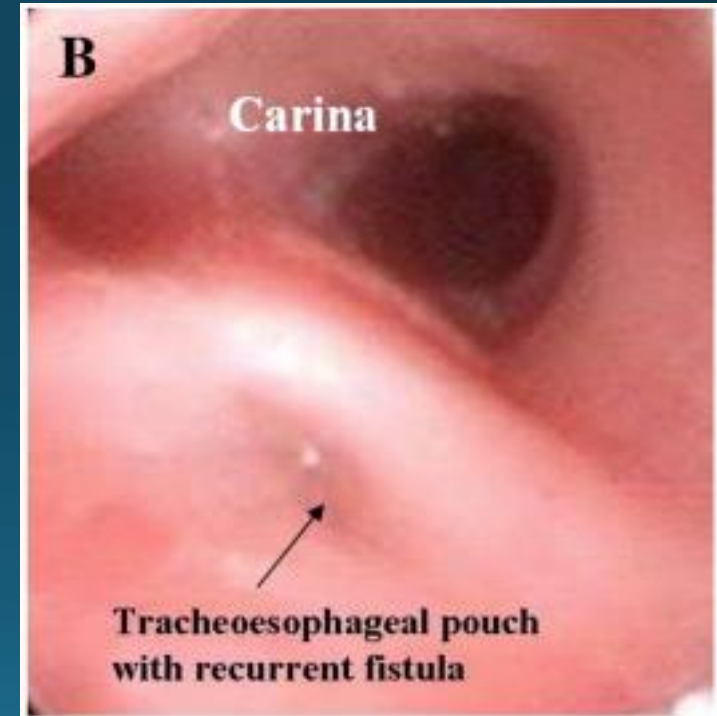
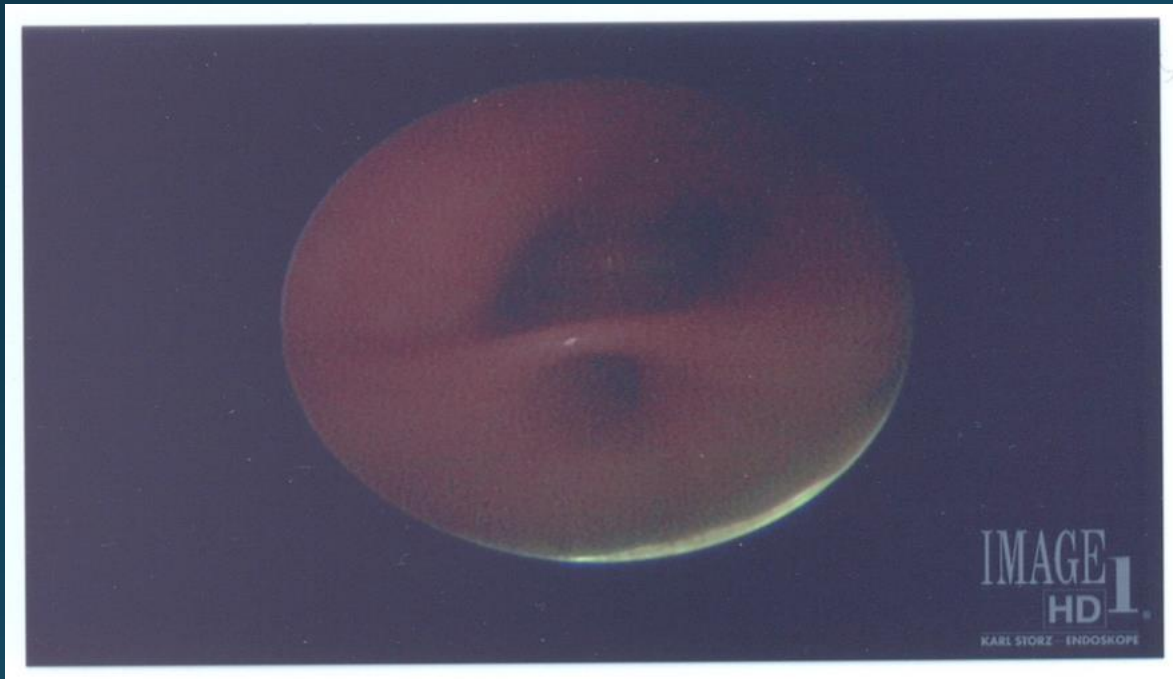
Presents for TEF repair

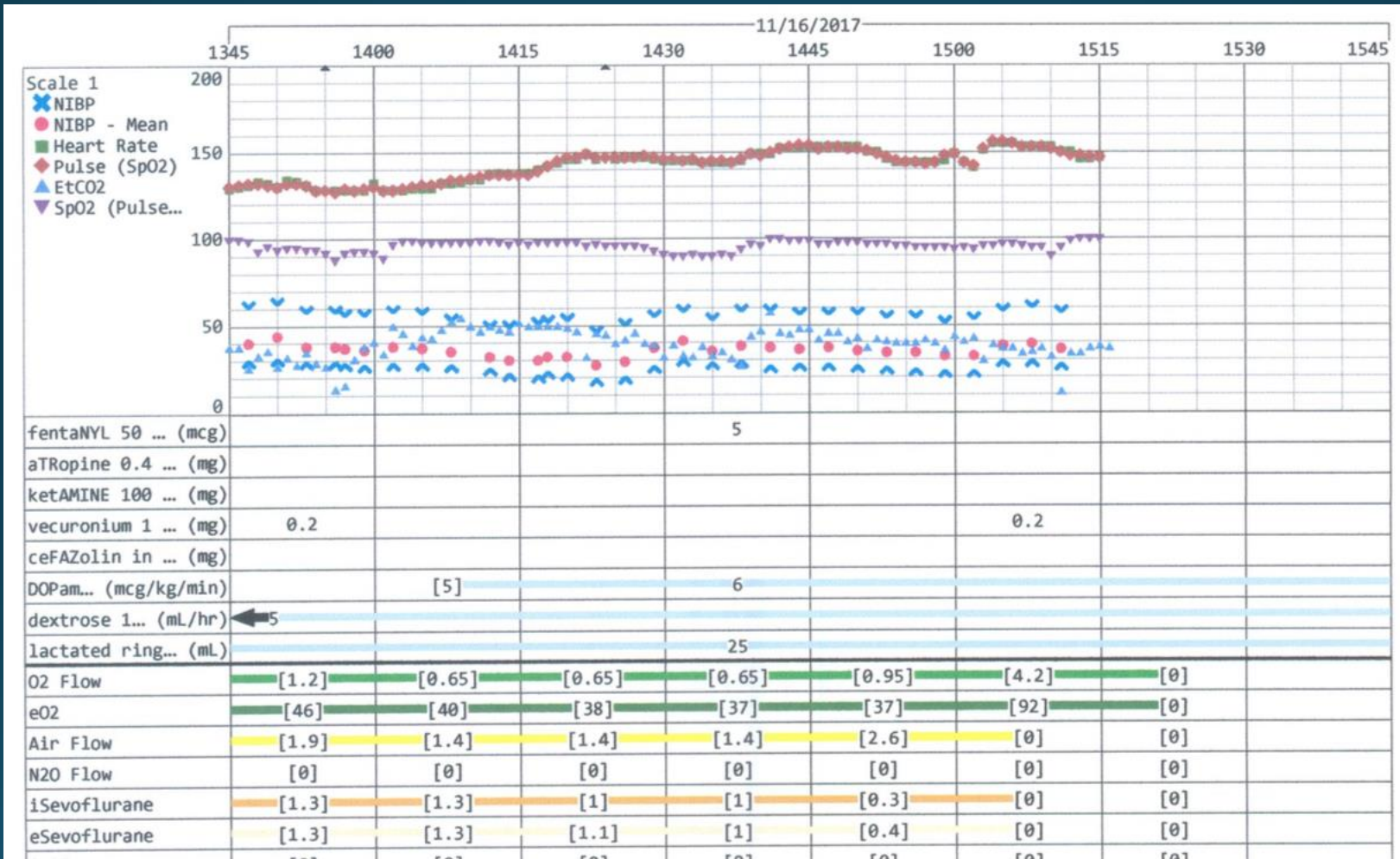
- ↑ respiratory support for ↑ WOB (2L)
- ↑ abdominal distension
- PIV x2, NGT in proximal esophagus
- CXR: Cardiomegaly with mild pulmonary edema
- SpO₂ intermittently 50%
- Blood and platelets transfused pre-op
- D10 @ 5 mL/hr
- Bidirectional shunting

Anesthetic Management - PREP

- Temp: warm room, underbody warmer, heat lamps
- Meds: dopamine, epinephrine gtt and IVP, phenylephrine, ketamine, NO
- Blood in room
- Airway: 3.0 ETT cuffed, additional sizes, LMA, CMAC, rigid bronchoscope
- Plan: target SpO₂ >80%, elective intubation by surgeon, if SpO₂ 60% begin NO, potential for palliative g-tube placement

Intraoperative Management





References

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