

Airway Management Challenge: Hunter's Disease

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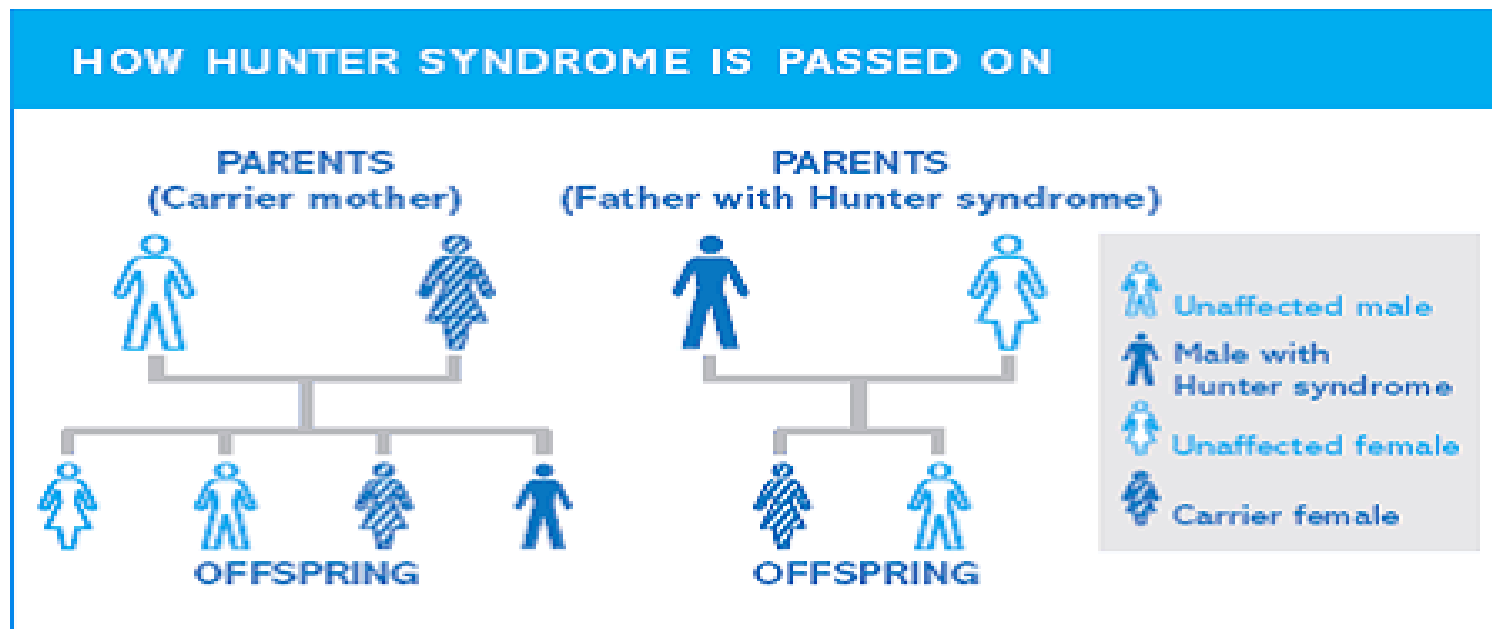
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BACKGROUND: Hunter's Disease



- Rare hereditary disease – 1:100,000 to 160,000
- X-linked recessive disorder primarily affecting males
- Deficiency or absence of the enzyme iduronate-2-sulfatase causing the build up of mucopolysaccharides (MPS), also known as glycosaminoglycans (GAG)
- Build-up of GAG results in physical manifestations and symptomatology
- Onset: 2-4 yrs old
- Always severe, progressive, fatal
- Life expectancy: <20 yrs old
- Death usually due to obstructive airway disease and/or cardiac failure

Inheritance Pattern: X-linked recessive



Hunter syndrome (MPS II) shows X-linked inheritance. On average, a carrier mother will pass on the abnormal I2S gene to 50% of her sons and 50% of her daughters. A father with Hunter syndrome will pass on the abnormal I2S gene to all of his daughters and none of his sons.

Signs and Symptoms



- developmental delay
- mental retardation
- short stature
- skeletal deformities
- enlarged tongue
- abnormal dentition
- sleep apnea
- upper airway obstruction
- decreased ROM
- large head
- valvular heart disease
- coronary disease
- myocardial infarction
- pulmonary hypertension
- hepatosplenomegaly
- enlarged abdomen
- hearing loss
- distinct facial features
- hydrocephalus
- increased intracranial pressure

Treatment

- Traditional: palliative focusing on management of clinical problems
- Current: enzyme therapy with idursulfase → recently approved by the FDA
- Enzyme therapy is not a cure yet improves the quality of life for children with Hunter's syndrome



Enzyme Therapy

- Researchers claim sustained **reduction** in urinary GAG excretion, liver/spleen size, heart size, and **improvement** in cardiac function, sleep apnea, joint mobility
- Reductions in urinary GAG excretion and size of the liver and spleen indicate that the enzyme is active and taken up into tissues and organs.
- Risks include infusion reactions such as cardiovascular instability, life-threatening respiratory depression, seizures
- Risks of implementing the treatment are minimal relative to the devastating course of the disease and lack of treatment

Case Study

- 13 y/o male with Hunter's disease diagnosed at age 2
- Weight: 22 kilograms
- Proposed Surgery: Tracheostomy
- Diagnosis: severe obstructive sleep apnea with excessive hypoxia and hypoventilation requiring BIPAP every night to maintain oxygen saturations above 95%
- Noncompliance w/ BIPAP due to mental status requiring restraints and resulting in injuries

Medical history

- Respiratory – increasing stridor and upper airway obstruction, recent aspiration pneumonia (resolved)
- Neurological – tonic-clonic seizures x1 per month, deteriorating mental status, cannot ambulate or speak
- Gastrointestinal – significantly delayed gastric emptying, PEG in situ

Cardiac status



- ECHO: LVDD, aortic/mitral valve thickening consistent with myxomatous malformation
- Cardiac anesthesiologist recommendations: Prophylactic antibiotics (subacute bacterial endocarditis), avoid propofol and tachycardia, assume pulmonary hypertension, maintain cardiac output and systemic vascular resistance

Current medications

- Albuterol, ipratropium bromide
- Budesonide
- Carbamazepine, Tegretol
- Cefotaxime, Clarithromycin, Erythromycin, Amoxicillin
- Lansoprazole
- Prednisone
- Idursulfase (enzyme therapy)



Pre-operative Exam

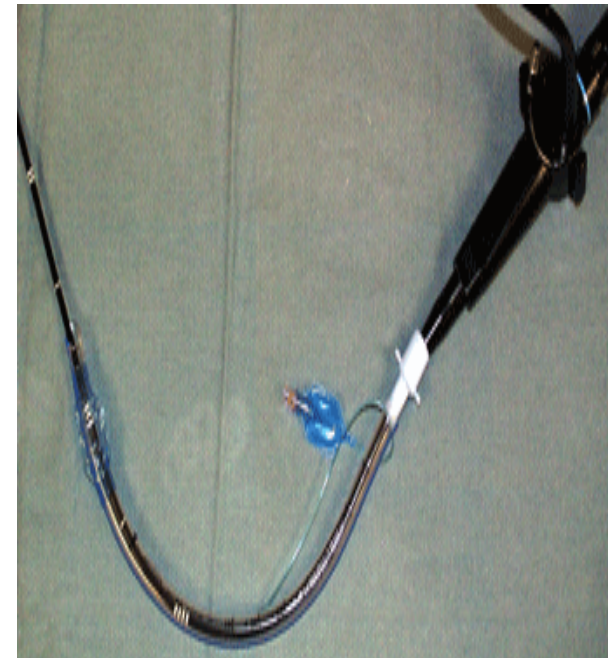
- Severe physical and developmental delay
- Micrognathia
- Short neck
- SMS – 3 cm
- Macroglossia
- Excessive salivation
- Mild wheezing and audible stridor
- Room air sat 94-96% w/ HOB 45 degrees

Anesthesia Plan

- ASA IV status
- Risks/benefits discussed with parents
- Emergency airway equipment, multiple LMAs, fiberoptic and rigid bronchoscopes, tracheostomy set-up
- Surgical team, two anesthesiologists, anesthesia technician, and SRNA present for induction

Anesthesia Plan...cont

- Maintain spontaneous ventilation
- Sevoflurane inhalational agent
- Fiberoptic bronchoscopy to intubate trachea UDVVC



Induction sequence

- Glycopyrrolate 0.22 mg IV
- Preoxygenation
- Stomach emptied via gastrostomy tube
- Mask w/ 100% oxygen and 2% Sevoflurane
- Sevoflurane increased incrementally to 3.2% while maintaining spontaneous ventilation

Induction...continued

- Loss of consciousness resulting in airway obstruction and decrease in oxygen saturation to 90%
- Oral airway alone did not alleviate obstruction
- Ventilatory assistance to maintain oxygen saturation >90% by one person providing jaw thrust, another maintaining tight mask seal, and a third delivering positive pressure
- Oxygen saturations were as low as 61% momentarily as ventilation was established

Intubation attempt



- DL X 2 by anesthesiologist and ENT surgeon revealing a severely obstructed view
- LMA placed requiring continued jaw thrust resulting in moderate air movement
- Fiberoptic introduced through LMA → tongue prevented advancement past the LMA
- LMA removed and mask ventilation as previously described

Intubation

- Tongue pulled forward
- Jaw thrust
- Fiberoptic bronchoscope advanced through left nare
- Grade IV view
- 5.0 ETT passed
- Confirmation of tracheal intubation
- Etomidate 2 mg, Fentanyl 10 mcg, Vecuronium 1.5 mg administered
- Hydrocortisone 50 mg, Ampicillin 1 gram given

Intraoperative

- Surgical procedure completed without incident
- Surgeon noted short neck and distorted anatomy resulted in a challenging and time consuming tracheostomy
 - thus an emergency tracheostomy would have been difficult

Intraoperative ... maintenance

- Cardiac considerations → Decreased volatile anesthetic concentrations by the titration of fentanyl, 100% oxygen
- Mechanical ventilation adjusted to maintain end tidal CO₂ 45-55 to reflect patient's baseline arterial carbon dioxide and, thus, elicit respiratory drive for spontaneous ventilation
- At the end of the case, Sevoflurane was turned off and 100% oxygen administered through the tracheostomy, patient was spontaneously ventilating and comfortable, ventilation was assisted by ambu bag to maintain oxygen saturation above 96%

Post-operative

- Transferred to PICU
- VSS
- Spontaneous ventilation through tracheostomy
- Post op Day 1 - Weaned from ventilatory assistance
- Post op Day 3- transferred to floor w/ resolution of obstructive sleep apnea and oxygen saturations >96% on room air

Discussion

- Hunter's syndrome is characterized by accumulation of MPS in vital organs
- MPS deposits in oral mucosa, epiglottis, tongue, tonsils, adenoids
- Abnormal tracheal and laryngeal anatomy
- Copious oral secretions

Discussion

- Respiratory tract changes begin from oral cavity and encompass supraglottic, epiglottic, and subglottic regions and extend to larynx, trachea and bronchioles → this patient was at the end of this continuum
- Neurological dysfunction results from the deposit of MPS in neurons of the respiratory center and in cranial nerve tracts leading to loss of pharyngeal wall tone and OSA

Discussion

- All of the above present the anesthesia professional with restrictive and obstructive ventilatory challenges
- Thorough pre-op evaluation should include consults w/ cardiac and pulmonary specialists
- ENT surgeons must be immediately available for possible emergency tracheostomy
- Preparation of equipment and trained personnel in anticipation of difficult ventilation and intubation
- Working knowledge of the difficult airway algorithm

Recommendation

- Maintain spontaneous ventilation
- Oral airway + jaw thrust + positive pressure ventilation to overcome obstruction
- FOB through LMA provides effective method for intubation as described in various case reports
- Induction and maintenance tailored to cardiac status, namely prevent tachycardia and hypotension

Conclusion

- Patients with Hunter's syndrome present complex airway management issues
- Research and clinical evidence emphasize inevitable difficult mask ventilation and tracheal intubation
- These patients become progressively challenging as MPS accumulates with age

Conclusion...cont



- Formulating a plan, seeking consultation of specialists, requesting assistance of colleagues and appropriate personnel, ensuring preparation of equipment, and discussing risks/benefits with caretakers are essential elements in the safe and effective care of patients with Hunter's disease

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