## Overview

- Craniofacial microsomia, also known as HFM or oculoauriculovertebral spectrum.
- Bilateral microsomia can occur.
- CNS, cardiac, and skeletal anomalies (expanded HFM spectrum) may occur
- Pulmonary, gastrointestinal, and renal deformities are less common.
- The majority of associated heart defects involve the outflow tract or septum. The increased frequency of cardiac anomalies with this condition suggests that abnormal development of the neural crest may result in both HFM and conotruncal heart defects.
- Children with HFM may have fused or hemivertebrae, resulting in limitation of neck flexion and extension and increasing the difficulty of intubation.
- There are positive correlations between the number of involved abnormal components and the degree of difficulty in visualizing the larynx in pts with both bilateral and unilateral microtia.
- Bilateral mandibular and auricular malformations increase the risk of difficult intubation.

## Etiology

- Rare congenital abnormality
- May result from chromosomal abnormality or disrupted blood flow to the head in utero

## Usual Treatment

- Removal of preauricular skin tags
- Remodeling, especially in the presence of orbital dystopia
- Orthodontic treatment
- Ear reconstruction
- Maxillary repositioning (Le Fort 1), mandibular advancement and soft tissue augmentation
- Mandibular distraction osteogenesis as required to facilitate subsequent intubations
- Rib grafting as required

## Assessment Points

| System | Effect | Assessment by Hx | PE | Test |
|---|---|---|---|---|
| HEENT | Mandibular hypoplasia | Facial asymmetry | Micrognathia, ear tags, OMENS | CT/MRI |
| CV | Conotruncal malformation | Dyspnea/poor feeding/delayed growth | Murmur | ECHO |
| MS | Fused or hem vertebrae | Neuromuscular changes | Limited neck flexion/extension | CT/MRI |

**Key References:** Nargozian C, Ririe DG, Bennun RD, et al.: Hemifacial microsomia: anatomical prediction of difficult intubation, *Paediatr Anaesth* 9(5):393–398, 1999; Walker RM, Ellswood J: The management of difficult intubation in children, *Paediatr Anaesth* 19(Suppl 1):77–87, 2009.

## Perioperative Implications

### Preoperative Preparation

- Conduct craniofacial assessment OMENS:
  - Orbit: Orbital distortion.
  - Mandible: Mandibular hypoplasia.
  - Ear: Microtia, periauricular tags.
  - Facial nerve: Facial muscle hypoplasia.
  - Soft tissue: Hypoplasia or absence of the parotid gland and masticatory muscles (temporalis, masseter).
- Review detailed history with surgeon, radiologist, and parents/guardians.
- Be prepared to call for help early.
- Assemble ear/nose/throat team in case rigid bronchoscopy or tracheostomy is required.
- Determine correct ET tube size and depth, as changing the tube or having too short a tube can lead to complications.
- Discuss and plan all approaches and backup plans.
- Check and prepare all the instruments before bringing the pt to the OR.
- Determine severity of mandibular hypoplasia in radiologic reports to estimate the degree of difficulty of intubation.
- Plan for difficult IV access. Presence of preexisting IV may facilitate concurrent IV and inhalational induction.

### Monitoring

- Arterial line may be required in the presence of cardiac or pulmonary morbidity.

### Airway

- Make your first attempt the best attempt.
- Assess and plan mask ventilation.
- For pts who are difficult to ventilate, oral, nasopharyngeal, or LMA insertion can improve ventilation.
- Avoid higher peak pressure as much as possible.
- If direct laryngoscopy fails, quickly go to video-assisted technique.
- Conventional laryngoscopy with a flat curved blade, such as a Macintosh, will be less helpful in a pt with micrognathia, as even the normal-sized tongue cannot be compressed adequately into the mandibular space to reveal the laryngeal structures.
- Paraglossal approach: Use a narrow, low-profile, straight-bladed laryngoscope in a paraglossal manner. Advance the blade in the space between the tongue and the lateral pharyngeal wall or tonsillar fossa.
- Lateral approach: The straight axis is shorter and insertion of the ET may be aided by a stylet or use of a gum elastic bougie. This approach is also called the retromolar approach, far lateral approach, and right molar approach.
- LMA can be used as a conduit for fiberoptic insertion of an ET tube.
- Confirm LMA placement with a fiberoptic examination.
- Use an antisialagogue to decrease airway secretions.
- It may be appropriate to leave the LMA in situ so as to minimize manipulation. Have a clear plan to remove it if necessary. Can load up two ET tubes back to back on the fiberoptic scope to facilitate removal of the LMA.
- Use humidified oxygen and steroid prophylactically after difficult intubation to minimize edema.
- Multiple approaches for laryngoscopy may be required.
- Direct laryngoscope with various blade sizes; McCoy and Sward laryngoscope blades.
- Video-assisted airway devices: Fiberoptic scope, Glidescope, Truview, etc.
- Supraglottic airways; LMA classic, Proseal LMA, iGel and COPA, etc.
- Hopkins rod rigid bronchoscope.
- Use Tegaderm/Vaseline gauze or hold the mask with both hands to improve seal.

### Postoperative Period

- Prolonged monitoring is recommended, especially when opioids are used to manage pain.
- Monitor pts for hemorrhage, regurgitation of gastric contents, hypoxic events.

## Anticipated Problems/Concerns

- Temporomandibular joint malformation may make jaw thrust difficult.

# Gonorrhea

Seth Eisdorfer

## Risk

- The prevalence of gonorrhea is decreasing, with 106.1:100,000 as of 2013.
- Most common in people ages 15–24 y, in large urban areas, and among people with low socioeconomic status and/or low levels of education.
- Incidence higher in men; prevalence higher in women.

## Overview

- Sexually transmitted disease.
- High incidence of coexisting chlamydial infection.
- Local infection: Purulent, profuse urethral discharge and possible epididymitis, prostatitis, or proctitis in men. It is often asymptomatic in women, but may have cervical discharge, vaginitis, salpingitis, or proctitis. Ascending infection may lead to PID.
- Disseminated infection: Fever/rash, tenosynovitis/arthritis (common), conjunctivitis (usually from autoinoculation), possible myopericarditis, and toxic hepatitis or perihepatitis (Fitz-Hugh-Curtis syndrome), rarely with endocarditis or meningitis.

## Etiology

- *Neisseria gonorrhoeae*: Gram-negative intracellular diplococcus, usually found inside polymorphonucleocytes.
- Humans only natural hosts for *N. gonorrhoeae*.

## Usual Treatment

- Dx gold standard involves the isolation of the organism by culture, testing for antimicrobial resistance.
- Test for other STDs, including syphilis and HIV; test partners as well.

- Penicillins and tetracyclines not recommended as first-line agents due to resistance.
- Fluoroquinolones no longer recommended as first-line therapy due to increasing resistance, especially in men who have sex with men.
- For uncomplicated cervicitis/urethritis, ceftriaxone is drug of choice; other third-generation cephalosporins (cefixime, cefpodoxime) are also commonly used. Spectinomycin can be used in penicillin allergic pts.
- Add doxycycline or azithromycin for coexisting chlamydial infections.
- Symptoms may subside without treatment, leaving a chronic asymptomatic carrier state.
- Pharyngeal infection is frequently asymptomatic; it may clear spontaneously over several wk, even without therapy. Ceftriaxone and trimethoprim-sulfamethoxazole can be used for treatment.
- Complicated infections can be treated via penicillin G IV × 5 d or ceftriaxone × 5 d. Oral fluoroquinolones may be used provided susceptibility.
- PID requires second-generation cephalosporin such as cefotetan or cefoxitin or a combination of clindamycin and gentamicin. Treat for chlamydial coinfection.
- Resolution of symptoms after treatment suggests cure; follow-up cultures are recommended.

## Assessment Points

| System | Effect | Assessment by Hx | PE | Test |
|---|---|---|---|---|
| HEENT | Conjunctivitis, ophthalmia neonatorum, adult gonococcal conjunctivitis | | | |
| | Pharyngeal infection | | Exudative tonsillitis | Cultures |
| GI | Anorectal infectionsProctitis | Pain, pruritus | Purulent discharge, bloody diarrhea | Cultures |
| GU | *Women* | | | |
| | Urogenital tract disease | Abnormal vaginal discharge, dysuria, urinary frequency, lower abdominal pain, labial pain, abnormal menstruation | Mucopurulent cervicitis | Cultures from urethra and vagina |
| | *Men* | | | |
| | Acute epididymitisProstatitis | Pain | | |
| CV | Gonococcal endocarditis | | Possible murmur | ECHO |
| GI | Perihepatitis (Fitz-Hugh–Curtis syndrome) | | RUQ tenderness | Liver enzyme elevation |
| GU | *Women* | | | |
| | PID | Lower abdominal pain, vaginal discharge, fever, palpable adnexal mass | Severe pain to palpation | Endocervix cultures |
| | *Men* | | | |
| | Urethritis | Dysuria | Purulent urethral discharge | Cultures from urethra |
| CNS | Gonococcal meningitis | | Meningeal signs | |
| MS | Septic arthritis | Most common cause of septic arthritis in young adults, tends to involve single joints | Warmth, tenderness of affected joint(s) | |
| DERM | Disseminated lesions | | | Ranging from maculopapular to pustular or hemorrhagic, usually peripheral |

**Key References:** Tapsall JW: *Neisseria gonorrhoeae* and emerging resistance to extended spectrum cephalosporins, *Curr Opin Infect Dis* 22(1):87–91, 2009; Centers for Disease Control and Prevention: Sexually transmitted disease surveillance, 2014. Atlanta, GA, 2015, US Department of Health and Human Services. <http://www.cdc.gov/std/stats>.

## Perioperative Implications

- Universal blood and body fluid precautions and/or barrier precautions

**Monitoring**

- Awareness of Foley catheter/temp probe placement

**Airway**

- Awareness if pharyngitis exists

**Positioning**

- Awareness of joint involvement

**Maintenance**

- Awareness of extent of disease

**Adjuvants**

- Vary with hepatic involvement.

## Anticipated Problems/Concerns

- No vaccine available
- Follow-up cultures
- Effective antibiotics
- Testing isolates for antibiotic susceptibility
- Routine culturing of high-risk populations
- Diligent contact tracing and prompt referral; treatment of sexual partners
- Education targeted at high-risk groups
- Use of condoms or other barrier methods

# Goodpasture Syndrome

Michael A. Hall | Lee A. Fleisher

## Risk

- Incidence of 1 case per million people per y.
- Accounts for 20% of cases of RPGN or crescentic glomerulonephritis.
- In terms of bimodal age distribution, more common in males 20–30 y of age and females 60–70 y of age.

## Perioperative Risks

- Anemia from recurrent or persistent intrapulmonary hemorrhage
- Hypoxia or hypoxic respiratory failure in cases of massive intrapulmonary hemorrhage
- Rapidly progressive renal failure or uremia
- Significant third-space fluid loss secondary to proteinuria

## Worry About

- Pts with active pulmonary hemorrhage may require mechanical ventilation in the postop period for hypoxic respiratory failure.
- Renal failure will alter drug pharmacokinetics and require adjustment of dosing or choice of anesthetic drugs.
- Anemia secondary to iron deficiency from repeated pulmonary hemorrhage, as well as anemia related to chronic kidney disease.
- Opportunistic infections such as pneumocystitis pneumonia in pts receiving immunosuppressive therapy.
- Volume overload in pts with severe renal insufficiency.

## Overview

- Rare, autoimmune, renal-pulmonary syndrome caused by autoantibodies directed against the glomerular basement membrane (anti-GBM antibodies).
- Major cause of RPGN, defined as a ≥50% loss of renal function (as quantified by glomerular filtration rate) over a 3-mo period.
- Usually presents with constitutional symptoms (night sweats, malaise), chronic cough progressing