Objectives:

- Recognize various diagnoses seen in pediatric plastic surgery
- Discuss various management options
- Determine appropriate referrals

Cleft/Craniofacial
- Congenital ear anomalies
- Head shape changes in infants
  - Deformational Plagiocephaly versus Craniosynostosis
- Cleft lip and palate
- Pierre Robin Sequence
- Unilateral Microtia
- Vascular Anomalies
  - Hand and Peripheral Nerve
    - Congenital hand anomalies
    - Hand trauma
    - Bacterial glove injuries
    - Facial palsy
  - Reconstructive
    - General wound and scar healing
    - Burns
    - Facial Fractures
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Congenital Ear Anomalies

- Most common: Stahl's ear, cup deformity or constricted ear, bat ear, lop ear, lidding, cryptotia, and rim kinks.
- Some require surgical repair when pt is older due to lack of skin and cartilage.
- Most of the above we can correct with Earmolds.
  - Should be placed prior to 7-10 days of age. We have an approximate 90% success rate.
  - For those few babies who do require surgery later, the deformity and the surgery are far less extensive when Earmolds have been used.
- Microtia
  - Sometimes associated with hemifacial microsomia

Congenital Ear Anomalies

- Earmolds
  - Dental clay is formed into a mold that can be customized to fit into any size helix (outer rim of the ear). Steri-strips, Mastisol adhesive, and Micropore tape are used to attach the molds to the part of the ear that is affected, which is then secured to your baby’s head. Sometimes Loctite superglue gel is also necessary.
Infant Head Shape Anomalies

Quick Anatomy Review

- 90% Adult Size by 1 year & 95% by 6 years

Terminology

- Normocephalic
- plagiocephalic: Non-specific term for head shape abnormality
- Brachycephalic: Flat posterior skull, may be epimetric or asymmete
- Dolichocephalic / Skaphocephalic: Long, skinny, tall head
- trigonocephaly: Forehead central prominence, "triangular"

Differential Diagnosis:

- Deformational (Positional) Plagiocephaly
  - Torticollis
- Craniosynostosis

Key Exam Points:

- Overall shape
- Suture Ridging
- bossing (area of skull prominence)
- fontanelle (bulging, size)
  - anterior: varies a lot in size/shape
  - posterior: smaller, closes sooner
- Ear position asymmetry
- Facial asymmetry (orbits, mid-face, mandible)

Deformational (Positional) Plagiocephaly

- Most common cause of infant head shape anomalies
- Typical pattern: severity frequently peaks at 4 months, then gradually improves

Risk Factors:

- Back-to-Sleep Campaign: Significant increase in incidence following 1990s push for positioning to reduce risk of SIDS
- Torticollis: Abnormal, asymmetric head or neck position commonly from SCM tightness
- Prolonged hospital stay
- Multiples
- Low muscle tone
- Hydrocephalus

What to advise:

- Reassurance: Head shape alone with adequate skull growth does not affect development or intelligence
- Reassurance: Most of the time this will resolve with time & growth
- Positional Education: Increase tummy time, alternate feeding sides, plastic when resting, alternate sides when sleeping
- Physical Therapy, if indicated for Torticollis. May require formal therapy or just educational visit to teach baby to change head position of autore position home exercise program
- Testing: XR is controversial. CT is very informative, but only indicated if there is a concern for craniosynostosis
- Refer if concerned

Craniosynostosis

Definition: Premature closure of 1 or more cranial sutures
Concerns: May cause intracranial pressure increase, may permanently distort facial features and head shape.

Types:

- Sagittal Synostosis: Premature closure of the sagittal suture
  - Most Common type (Over 50%)
- Coronal Synostosis: Frontal boss asymmetry
  - High arching orbit (Harlequin feature)
- Metopic Synostosis: Normally does not affect others. May cause a "pointed" or "triangle" shape to the forehead.
  - Common to have a bony ridge or prominence in the forehead that will correct without surgical intervention.
- Lambdoid Synostosis: Often difficult to differentiate, flat posterior with a mastoid bulge and lower face asymmetry

When to refer to a specialist:

- Head shape worsening instead of improving with time
- Head shape changes present and unchanged since birth
- Suture ridging
- Significant facial asymmetry
- Parental Concern
- Unresponsive to conservative measures for 2-3 months or by 6 months of age
Cleft Lip and Palate

- Most common congenital anomalies of the face and skull, affecting approximately one in 700 newborns in the U.S.
- Both the lip and palate affect about 50% (M>F and L side>R side)
- Isolated cleft palate alone affects about 30% (F>M)
- Cleft of the lip alone affects about 20%

* The rates do vary by ethnicity with higher rates (one in 500) in patients of Asian descent and lower rates (one in 2,000) in patients of African descent.

Unilateral Cleft Lip

Bilateral Cleft Lip
Cleft Lip and Palate

- Typical sequence and timing of clinic visits and potential surgeries needed for patients with cleft lip and palate:
  - Weight Check and Feeding Clinic with Plastic Surgery PA and Speech Therapist
  - NAM therapy with Orthodontist
  - ENT to eval for ear tubes to be placed during simultaneous surgery with PRS
  - Meet Craniofacial Surgeon
  - Cleft lip repair (age 3 to 4 months)
  - Meet Cleft/CRF Team and return annually
  - Cleft palate repair (age 9 to 12 months)
  - Meet Speech Therapy needs
  - Possible revision surgery for speech issues related to cleft palate (age 4 and older)
  - ABG: Bone graft from the hip to the alveolus (ages 7 to 10)
  - Possible surgery to align the jaws once growth is complete (ages 16 to 18)
  - Possible revision to the nose or lip usually after a decision about jaw surgery is made

Pierre Robin Sequence

- Occurs in approximately one in 8,500 to 14,000 births, equally as common in males and females
- When the following birth defects occur together:
  - Micrognathia – Small lower jaw
  - Glossoptosis – Downward or backward positioned tongue
  - Breathing and feeding difficulty
  - +/- Cleft palate
- Sequence, not a syndrome, because it begins as a sequence of events:
  - underdeveloped lower jaw → abnormal displacement of the tongue → formation of a cleft palate
- Unknown cause, may be a part of many genetic syndromes
- Genetic testing recommended to determine if pt has any associated anomalies (i.e. Stickler Syndrome, Velo-Cardio-Facial Syndrome)
- Treatment
  - Conservative: Prone positioning
  - Surgical: Tongue stitch, Distraction Osteogenesis

Hemifacial Microsomia

- 2nd most common congenital facial anomaly (behind cleft lip/palate)
- Affects about 1 out of every 3500-4000 live births
- Congenital condition in which the tissues on one side of the face are underdeveloped.
- Synonyms include: craniofacial microsomia, first and second branchial arch anomaly, branchial arch syndrome, facioauriculovertebral syndrome, oculoauriculovertebral spectrum, and lateral facial dysplasia
- Cause is unknown. Research has shown the process starts in the first trimester of pregnancy and may be caused by a vascular problem leading to poor blood supply to the fetus’ face during early development.
- Typically not inherited. In a small minority of cases, a child may inherit the condition from his parents. An adult with hemifacial microsomia has about a 5% chance or less of having a child with the same condition.
Hemifacial Microsomia

- **OMENS Classification System**
  - **O**: orbit
    - O(0): normal size and position
    - O(1): abnormal size
    - O(2): abnormal position
    - O(3): abnormal size and position
  - **M**: mandible
    - M(0): normal
    - M(1): mandible small with short ramus
    - M(2): mandibular ramus is short and abnormally shaped
      - 2A: glenoid fossa in acceptable position
      - 2B: TMJ is displaced with severely hypoplastic condyle
    - M(3): complete absence of ramus, glenoid fossa, and TMJ
  - **E**: ear
    - E(0): normal
    - E(1): mild hypoplasia and cupping with all structures present
    - E(2): absence of EAC with variable hypoplasia of concha
    - E(3): malpositioned lobule with absent auricle, lobule remnant usually inferiorly and anteriorly displaced
  - **N**: nerve
    - N(0): no nerve involvement
    - N(1): upper facial nerve involvement
    - N(2): lower facial nerve involvement
    - N(3): all branches of facial nerve involved
  - **S**: soft tissue
    - S(0): no obvious deficiency
    - S(1): minimal deficiency
    - S(2): moderate deficiency

Treatment of Hemifacial Microsomia

- Treatment is aimed at correcting/reconstructing affected areas.
- Treatment for the mandible can include orthodontics, distraction osteogenesis, reconstruction with rib graft or free fibula graft.
- Treatment of ear abnormalities (microtia) includes ear reconstruction with rib cartilage graft vs reconstruction with implant such as Medpor.
- Treatment for the soft tissue can include fat grafting.
- Treatment for orbital abnormalities can include a variety of reconstruction procedures.
- Treatment for nerve abnormalities can include facial reanimation surgery.

Cleft/Craniofacial Team

- The Cleft and Craniofacial Team at Children’s Health is proud to be an ACPA (American Cleft Palate-Craniofacial Association) -designated team.
- Our weekly team visits include these specialties:
  - Plastic surgery
  - Oromaxillofacial surgery
  - Otolaryngology
  - Psychologist
  - Pediatric Dentist
  - Orthodontist
  - Speech therapist
  - Social worker
  - Geneticist
Vascular Anomalies

All inclusive term that includes vascular malformations and vascular tumors.

Vascular malformations
- Venous malformations
- Lymphatic malformations
- Arteriovenous malformations
- Capillary malformations (Port Wine Stain): made up of small blood vessels inside the skin and commonly occur on the face, but can be found on other parts of the body.
- Treat with laser treatments every 6-8 weeks.

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Vascular Malformations

- **Lymphatic Malformations**: large, soft masses commonly in the head and neck region.
  - Diagnosed by physical exam and MRI
  - Treat with sclerotherapy by IR or surgical excision

- **Venous Malformations**: usually cause no symptoms and require no treatment unless symptomatic.
  - Diagnosed with physical exam and MRI
  - Treat with compression garments, sclerotherapy or surgical debulking

- **AV Malformations**: abnormal connection between arteries and veins.
  - Diagnosed by physical exam (bruit, thrill) and MRI
  - Treat with embolization and radial surgical removal

Vascular Tumors

- **Hemangioma**: benign tumor of dilated blood vessels.
  - Most common tumor of infancy, 1 in every 10 children is affected. 32-34% of premature infants will have a hemangioma.
  - Present shortly after birth as a flat, red spot and may grow until 6-12 months of age.
  - Involution (shrinking) phase usually begins at 18-24 months. Process can last up to 5 years.
  - Treatment: time, propranolol or timolol, laser, surgery
Propranolol for Hemangiomas

- Children's Medical Center Propranolol Protocol
  - Indications for treatment: risk of functional impairment (periocular or perioral), rapid growth, ulcerations, risk of disfigurement
  - Baseline evaluation: birth history, vital signs, medical history
  - Initiation: Propranolol 10mg/5mL oral suspension
    - 1 mg/kg/day divided three times daily (every 8 hours)
    - If under 2 months corrected age or below 1% for weight
    - Inpatient admission to Hematology service for initiation
    - If over 2 months corrected age, safe for outpatient initiation
  - Follow-up
    - Follow-up 1 week for VS check and increase of dose to 2mg/kg/day divided three times daily (treatment dose)
    - Continue to follow monthly for VS check and dose adjustments for weight gain
  - Cessation of treatment
    - Usually after 12 months of treatment (treat through entire proliferative phase)
    - Decrease dose by half for 1 week then discontinue.
- In 2014, Propranolol (Hemangeol) was FDA approved for the treatment of infantile hemangiomas.

Vascular Anomalies Team Clinic

- Children with complex vascular anomalies are managed at Children's Medical Center in a team fashion which includes:
  - Plastic Surgeon
  - Dermatologist
  - Otolaryngologist
  - Hematologist
  - Interventional Radiologist

Hand and Peripheral Nerve

- Congenital hand anomalies
- Hand trauma
- Brachial plexus injuries
- Facial palsy
Congenital Hand Anomalies

- **Polydactyly**
  - Radial: thumb duplication. Surgical reconstruction
  - Ulnar: Type A is well formed and needs surgical removal. Type B are skin tags and can be clipped in nursery or in first few weeks of life

- **Syndactyly**
  - One of the most common, 1 per 2,000-2,500 life births, 3rd webspace is most common
  - Associated with Poland or Apert Syndrome
  - Can be skin only (simple) or include bone (complex)

- **Camptodactyly**
  - Congenital flexion deformity of PIP joint, SF most common
  - Splinting may be attempted to correct deformity. Surgery if needed

- **Clinodactyly**
  - Curvature of a digit in a radioulnar plane. Most common is SF DIP joint. Treatment is not usually necessary unless causing functional problems

- **Trigger Thumb**
  - Inability to extend at IP joint, palpable nodule over flexor MCP joint
  - Spontaneous resolution can occur in 1/3 of children by 1 year of age
  - Surgery recommended before 3 years of age, division of A1 pulley

Basics of Pediatric Hand Trauma

- **Fractures** – Treat the patient, not the Xray
  - Salter Harris Scale
  - Bottom Line: Non-displaced, no scissoring, no extensor lag, no malrotation = place in splint, followed by casting for 4-6 weeks of immobilization

- **Lacerations** – Bottom Line: Use absorbable sutures!
  - Most common injury = finger tip/nail bed injury +/- tuft fracture
  - Always always ‘shut in doors’
  - Subungual hematoma = fracture = technically an open fracture
  - Remove nail plate, repair nail bed, then replace nail stent
  - If more of the fingertip is involved, suture tip in place to act as a biologic dressing
  - Immobilization crucial for healing/regeneration
  - Traumatic amputation requiring replant
  - Bottom Line: Success is not measured by survival of the replanted tissue, but by the extent of return of useful function. Function anticipated after replantation should be better than that expected with amputation and prosthesis.
Brachial Plexus Injuries

- Brachial plexus birth palsy is an injury to the brachial plexus nerves that occurs during childbirth. The nerves of the brachial plexus may be stretched, compressed, or torn in a difficult delivery. The result might be a loss of muscle function, or even paralysis of the upper arm. Injuries may affect all or only a part of the brachial plexus.
  - Brachial plexus birth palsies occur in about one to three out of every 1,000 births.
  - Risk factors for sustaining brachial plexus birth palsy include: large gestational size, breech birth, prolonged or difficult labor, vacuum-or forceps-assisted delivery, twin or multiple pregnancy, history of a prior delivery resulting in brachial plexus birth palsy.
  - Traumatic brachial plexus injury can occur at any age—often as a result of a sports injury or car or work accident.

Brachial Plexus Birth Injuries - Treatment

- Observation: Most brachial plexus birth palsies will heal on their own. Your doctor will monitor your child closely. Many children improve or recover by 3 to 12 months of age. During this time, ongoing exams should be performed to monitor progress.
- Physical therapy and/or occupational therapy: Therapy is recommended to help maximize use of the affected arm and prevent tightening of the muscles and joints. With the teaching and guidance of therapists, parents learn how to perform range of motion (ROM) exercises at home with their child several times a day. These exercises are important to keep the joints and muscles moving as normally as possible.
- Botulinum toxin injections: May be used (mainly for the shoulder) to help with joint motion, rebalancing muscles, preventing contractures and shoulder dislocations.
- Surgery: Children who continue to have problems three to nine months after birth may benefit from surgical treatment. There are several surgical options, including microsurgery to repair or reconstruct the injured nerves, tendon transfers, muscle transfers, or joint surgery.

Facial Palsy

- Congenital versus acquired
- Complete, incomplete, or no recovery
- Main concern is eye protection
- Surgical consideration
  - Early upper eyelid intervention if needed
  - Facial reanimation—focused on restoring smile
    - Age 4-5 in congenital cases
    - After 18 months of plateau in improvement of acquired cases
  - One versus two stage procedures
Facial Palsy

- One stage procedure:
  - Microsurgical transfer of muscles to nerves that normally control the biting motion.
  - The ability to smile can be restored, but the patient must bite in order to activate the muscles.
  - If both sides of the face are involved, this is usually done one side at a time.
- Two stage procedure:
  - The first stage involves transferring nerves from "the good side of the face" to the paralyzed side of the face.
  - Then, nine months later, a muscle transfer using microsurgical technique is performed to the previously transferred nerves. If this is successful, nerve activity from "the good side of the face" travels instantly through the transferred nerves to the new muscle on the opposite side. This can return the ability to spontaneously smile. However, the smile motion is, at best, unrefined because a few nerves and one muscle are being asked to take the place of many muscles that work together during normal facial expression.

Reconstructive Surgery

- General wound and scar healing
- Burns
- Facial Fractures
- Other reconstructive procedures

Wound Healing/Scar Care

- Scars can take up to one year to completely mature. We do not consider scar revision until 1 year or more from injury.
- At least twice daily application of mild, fragrance and dye free moisturizer (cetaphil, eucerin, aquaphor).
- Strict sun avoidance: high SPF re-applied every 30 minutes while outside.
  - Translucent titanium or zinc oxide are ideal: provide physical barrier
- Silicone gels/sheeting have some literature support
  - Good clinical outcomes in our experience
- We do not recommend other commercial scar care products
- Scar massage if tolerated for thick, firm subcutaneous scar tissue.
Burns

<table>
<thead>
<tr>
<th>Degree</th>
<th>Depth</th>
<th>Appearance</th>
<th>Sensation</th>
<th>Time to Healing</th>
</tr>
</thead>
<tbody>
<tr>
<td>First</td>
<td>Confined to epidermis</td>
<td>Erythema. Does not blister initially, and usually sloughs off.</td>
<td>Dry, Mild to moderate sensation.</td>
<td>Few days.</td>
</tr>
<tr>
<td>Third</td>
<td>Entire epidermis and dermis</td>
<td>Stiff white or tan. Do not blanche.</td>
<td>Dry, Charred, Insensate</td>
<td>Skin graft required. May need escharotomy.</td>
</tr>
</tbody>
</table>

Surgical management of burns:
- First goal is to restore function and protection of vital structures, then the aesthetic appearance
- Urgent procedures are for burns that involve exposed or severely damaged structures
- Early procedures are to improve burn care and rehabilitation and non-vital function
- Late procedures include those with a goal of best aesthetic result possible
- Examples of surgical care: primary closure, excision and skin graft, skin substitutes, tissue expansion, tissue transfer (local, distant and free flap)

Pediatric Facial Fractures

Most Common Causes:
- Falls
- Athletics
- Physical Aggression
- MVC/MVA

Common Facial Fractures:
- Nasal: Most Common: Nasal swelling and tenderness +/- deviation
- Orbital: Periorbital bruising, tenderness, eye swelling and difficulty with motion
- Nasoorbitoethmoid (NOE): telecanthus, enophthalmos, diplopia, and apparent midface retrusion.
- Mandible: Jaw Dislocation: Inability to open/close jaw

Key:
- Most are non-surgical
- Cranial Volume > Facial Volume
- Increased fat pad coverage in children
- Bones malleable

Key:
- Jaw Dislocation: Inability to open/close jaw

Exam:
- Overall: Facial symmetry
- Skin: Bruising Patterns, Lacerations
- Eye: Motions, Orbital Examinations
- Nasal: Deviation, Septal Hematoma
- Skin: Intra-oral: Tongue blade bite test
- Intermaxillary: Dental Fractures, bleeding oral turfs
Facial Fractures: When to refer

Key: Again – Most are non-surgical

Emergent
- Orbital entrapment - any concern for eye motion - You must assess eye motion!
- NOE - Any sign of increased intercanthal distance, enophthalmos or mid face retrusion
- Nasal Septal Hematoma - yes, but will need to be drained to reduce risk of septal perforation, abscess formation and saddle nose deformity
- Open Mandible fracture – concerns same with present of fracture with tooth fracture / bleeding

Urgent
- Nasal (Displaced) - prefer to reduce at 5-7 days
- Mandible (closed, displaced) - mild occlusion, difficulty opening/closing

Non-urgent facial fracture management
- Soft diet
- Gentle motion
- Limit risky activities
- Nasal: No nose blowing, nasal decongestant

Consults to consider:
- Dental
- Ophthalmology
- Neurology

Thank you!