Craniofacial Anomalies and coding from common to rare

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Craniofacial Anomalies

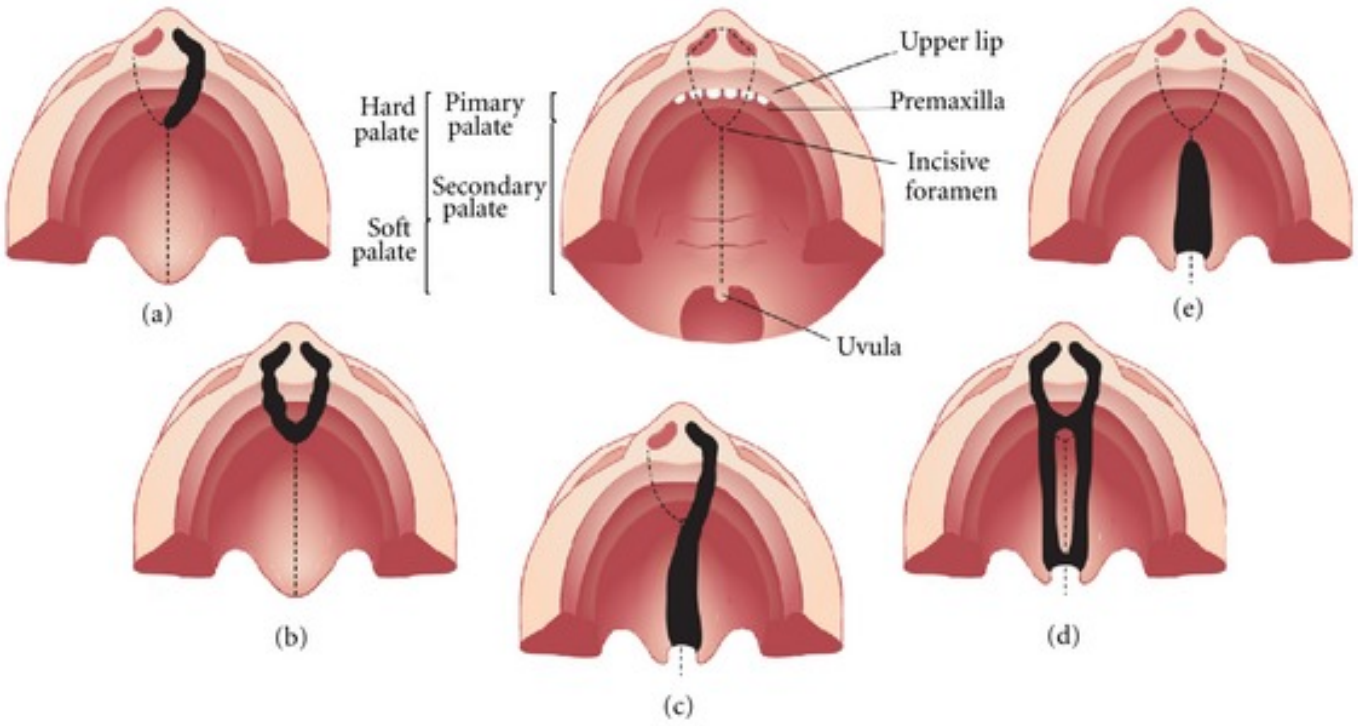
Common

- Cleft lip and/or Cleft palate
- Facial Palsy
- Craniofacial Fibrous Dysplasia
- Hemangioma

Rare

- Antley-Bixler Syndrome
- Craniosynostosis
- Crouzon syndrome
- Goldenhar Syndrome/Hemifacial microsomia
- Treacher-Collins Syndrome

Cleft lip and/or Cleft palate
-Can be found on Ultrasound



- **Figure 1**: Representation of the most common types of cleft affecting the palate.
- (a) Unilateral cleft lip with alveolar involvement
- (b) bilateral cleft lip with alveolar involvement
- (c) unilateral cleft lip associated with cleft palate
- (d) bilateral cleft lip and palate
- (e) cleft palate only

Cleft lip and/or Cleft palate

Notations

- Use Additional code to identify associated malformation of the nose (Q30.2)
- Cleft Palate: Includes
  - fissure of palate
  - Palatoschisis
- Cleft Lip: Includes
  - cheiloschisis
  - congenital fissure of lip
  - harelip
  - labium leporinum
- Cleft Palate and Lip: Includes
  - cheilopalatoschisis

Signs, Symptoms and Complications

- Feeding Difficulties

  Under 28 days:

  P92.8 Other feeding problems of newborn

  Over 28 days:

  R63.3 Feeding difficulties

Cleft lip and/or Cleft palate

Diagnoses

- Q35.1 Cleft hard palate
- Q35.3 Cleft soft palate
- Q35.5 Cleft hard palate with cleft soft palate
- Q35.7 Cleft uvula
- Q36.0 Cleft lip, bilateral
- Q36.1 Cleft lip, median
- Q36.9 Cleft lip, unilateral
- Q37.0 Cleft hard palate with bilateral cleft lip
- Q37.1 Cleft hard palate with unilateral cleft lip
- Q37.2 Cleft soft palate with bilateral cleft lip
- Q37.3 Cleft soft palate with unilateral cleft lip
- Q37.4 Cleft hard and soft palate with bilateral cleft lip
- Q37.5 Cleft hard and soft palate with unilateral cleft lip
- Q37.8 Unspecified cleft palate with bilateral cleft lip
- Q37.9 Unspecified cleft palate with unilateral cleft lip
- Q30.2 Fissured, notched and cleft nose
- K00.1 Supernumerary teeth
- K00.2 Abnormalities of size and form of teeth
- K00.3 Mottled teeth
- K00.4 Disturbances in tooth formation
- K00.5 Hereditary disturbances in tooth structure, not elsewhere classified
- K00.6 Disturbances in tooth eruption

Cleft lip and/or Cleft palate

Procedures

- 14040 Adjacent tissue transfer or rearrangement, forehead, cheeks, chin mouth, neck, axillae, genitalia,
- hands and/or feet; defect 10sq cm or less
- 14041 defect 10.1 sq cm to 30.0 sq cm.
- 14060 Adjacent tissue transfer or rearrangement, eyelids, nose, ears, and/or lips; defect 10 sq cm or less
- 14061 defect 10.1 sq cm to 30.0 sq cm
- 15120 - 15261 (additional reconstructive codes under the Integumentary System)
- 15576 Formation of direct or tubed pedicle, with or without transfer; eyelids, nose, ears, lips, or intraoral
- 20902 Bone harvest any area, major or large.
- Code used for obtaining autogenous bone or other tissues through a separate skin incision by a separate surgeon than performing the primary procedure. If the primary procedure “includes obtaining the graft” use the -52 modifier on the primary procedure for reduced services or both surgeons may report the primary procedure code appended by the -62 modifier.

Cleft lip and/or Cleft palate

- For example: surgeon #1 repairs a unilateral cleft: use 42210-52.
- Surgeon #2 harvest bone from the iliac crest and does not assist in the surgery: use 20902-62

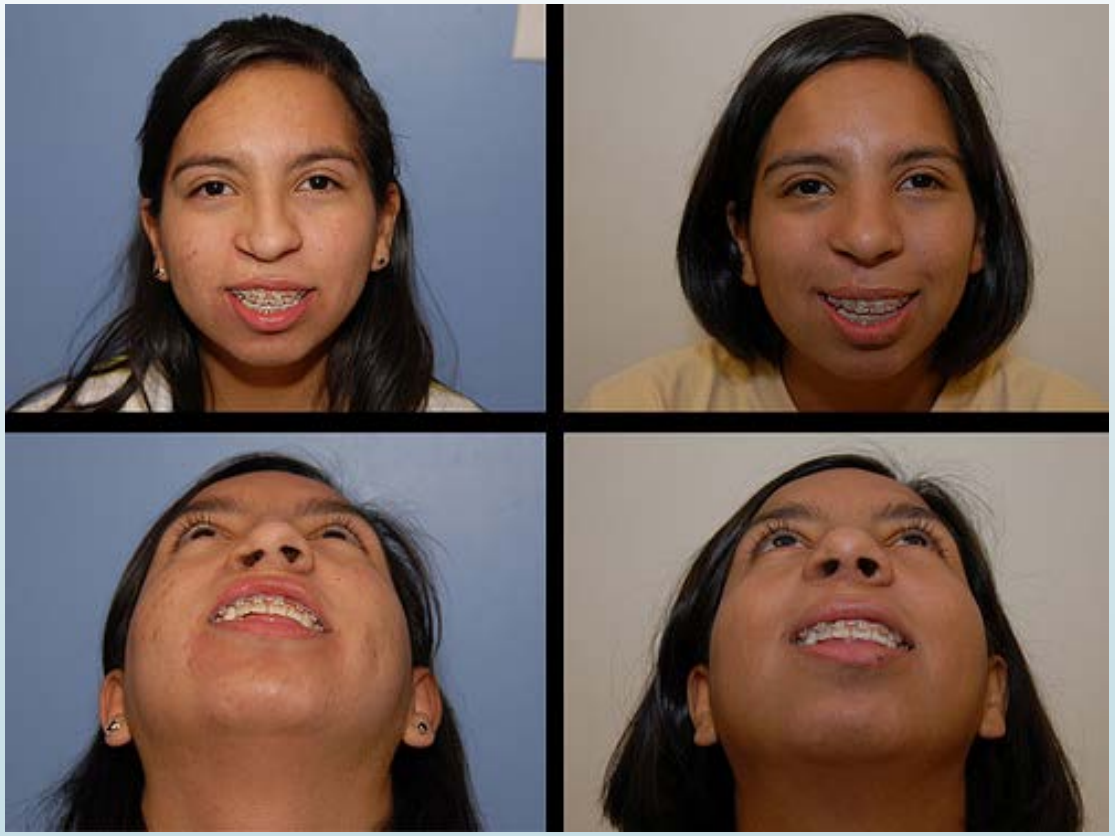
- 30580 Repair fistula; oromaxillary (combine with 31030 if antrotomy is included)
- 30600 Repair oral-nasal fistula
- 30400-30630 Rhinoplasty, septoplasty, nasal region repair codes
- 40650 Repair lip, full thickness; vermillion only
- 40652 Repair Lip, full thickness, up to half vertical height
- 40654 Repair Lip, full thickness over one-half vertical height, or complex

Cleft lip and/or Cleft palate

- 40700 Plastic repair of cleft lip/nasal deformity; primary, partial or complete, unilateral
- 40701 primary bilateral, one stage procedure
- 40702 primary bilateral, one of two stages
- 40720 Secondary repair of cleft lip/nasal, by recreation of defect and reclosure
- 40761 with cross lip pedicle flap (Abbe-Estlander type), including sectioning and inserting pedicle
- 42200 Palatoplasy for cleft palate, soft and/or hard palate only
- 42205 Palatoplasty for cleft palate, with closure of alveolar ridge; soft tissue only
- 42210 Palatoplasty for cleft palate with bone graft to alveolar ridge (includes obtaining graft). This includes grafting from the alveolar crest to the piriform rim.

Cleft lip and/or Cleft palate

- 41899 Extraction of teeth during cleft palate surgery.
- 42215 Palatoplasty for cleft palate; major revision
- 42220 Palatoplasty for cleft palate, secondary lengthening procedure
- 42225 Palatoplasty for cleft palate, attachment pharyngeal flap
- 42226 Lengthening of palate and pharyngeal flap
- 42227 Lengthening of palate, with island flap
- 42235 Repair of anterior palate, including vomer flap
- 42260 Repair of nasolabial fistula



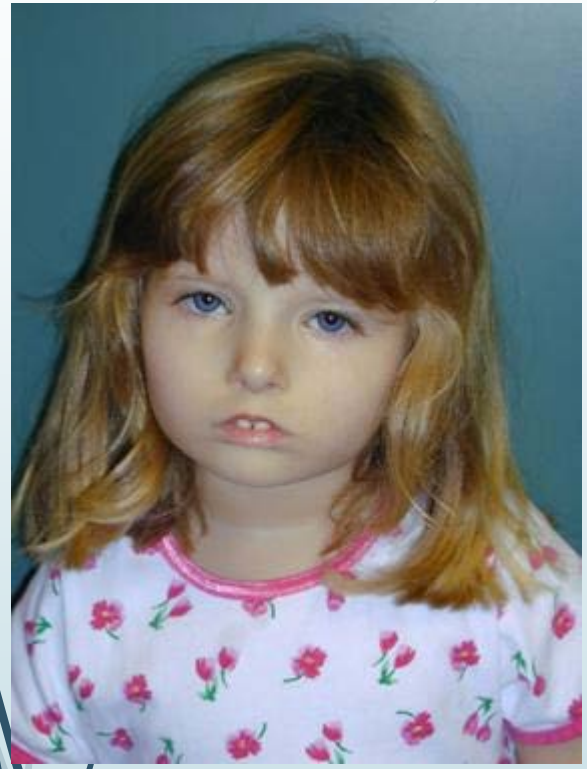
ICD-10-PCS - Cleft lip and/or Cleft palate

- 0JX---- Transfer, Subcutaneous Tissue and Fascia
- 0HRX-- Replacement, Skin and Breast
- 0H0--- Alteration, Skin and Breast=Skin Substitute Grafts
- 0HW--- Revision, Skin and Breast=Revision of Skin Grafts
- 0HX--- Transfer, Skin and Breast=Autologous Skin Grafts
- 0QS---- Reposition, Lower Bones=Bone graft
- 0PS---- Reposition, Upper Bones=Bone graft
- 0NU---- Reposition, Head and Facial Bones=Bone graft

ICD-10-PCS - Cleft lip and/or Cleft palate

- **0CQ00ZZ Repair Upper Lip, Open Approach**
- **0CQ01ZZ Repair Lower Lip, Open Approach**
- **0CQ02ZZ Repair Hard Palate, Open Approach**
- **0CQ03ZZ Repair Soft Palate, Open Approach**
- **09QK0ZZ Repair Nasal Mucosa and Soft Tissue, Open Approach**
- **0CDWXZ- Extraction, Upper Tooth**
- **0CDXXZ- Extraction, Lower Tooth**

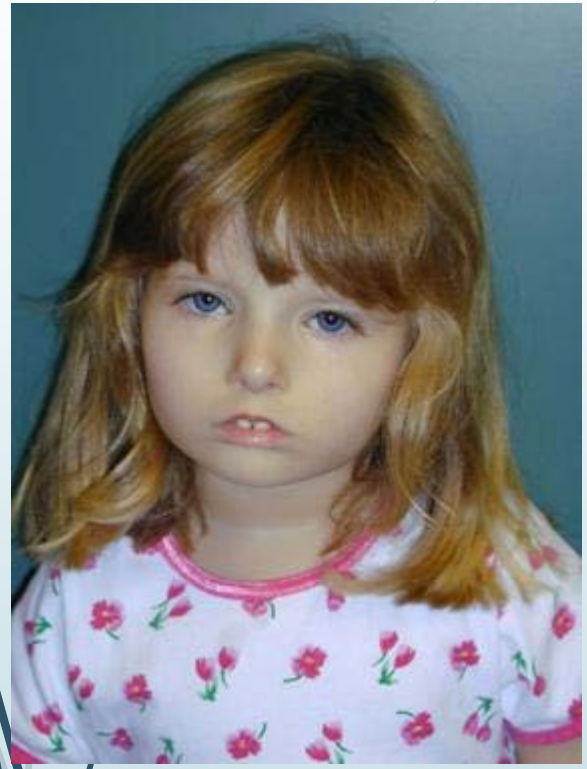
Congenital Facial Palsy-What is it?



Möbius syndrome

- Congenital (present at birth) facial paralysis is uncommon and, when present, may cause multiple problems for the newborn, such as difficulty with nursing and incomplete eye closure. If the paralysis does not resolve, it may affect the child's future speech, expressions of emotion, and mastication. This article discusses the etiologies, evaluation, diagnostic testing, and treatment options for this disorder. [1]
- Congenital facial paralysis is classified as traumatic or developmental, unilateral or bilateral, and complete or incomplete (paresis). Determining the etiology is important because the prognosis and treatment differ, depending on the underlying pathophysiology. An appropriate history and physical examination usually resolve the origin, but radiographic imaging and neuromuscular testing may be necessary for treatment planning

Congenital Facial Palsy-What is it?



Möbius syndrome

- Conversely, no procedures are available that can enable an infant to develop normal function of the facial nerve when the palsy is developmental in origin. Facial reanimation's goal is to minimize asymmetries and improve function. Surgical exploration in the newborn with facial paralysis is controversial. Issues regarding timing of facial rehabilitation are complex. The factors that are involved include ability of the infant to tolerate a surgical procedure, the unknown potential for recovery, and whether early surgical intervention can prevent future psychosocial problems for the child.

Congenital Facial Palsy

Causes

- Trauma
- Möbius syndrome=Q67.0
- Hemifacial microsomia-=Q67.0
- 22q11.2 deletion syndrome (22qDS)= Q93.81
- Albers-Schönberg disease=Q78.2
- CHARGE syndrome=Q30.0
- Facioscapulohumeral muscular dystrophy=G71.0
- Congenital unilateral lower lip paralysis/asymmetric crying faces=Q18.6
- Teratogens=Q99.8

Congenital Facial Palsy

Codes

- Q07.8=Other specified congenital malformations of nervous system
  - Agenesis of nerve
  - Displacement of brachial plexus
  - Jaw-winking syndrome
  - Marcus Gunn's syndrome
- Q67.0=Congenital facial asymmetry
  - Acrocephalopolysyndactyly
  - Acrocephalosyndactyly [Apert]
  - Cryptophthalmos syndrome
  - Cyclopia
  - Goldenhar syndrome
  - Moebius syndrome
  - Oro-facial-digital syndrome
  - Robin syndrome
  - Whistling face
- P11.3=Birth injury to facial nerve

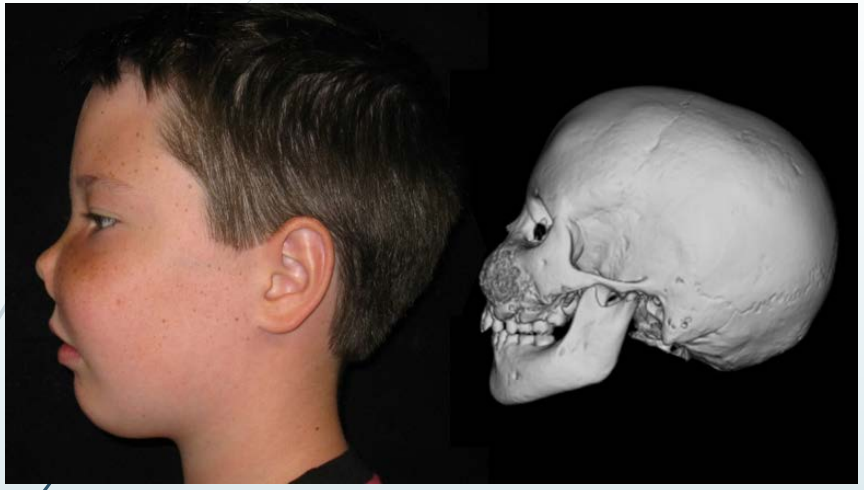
Congenital Facial Palsy

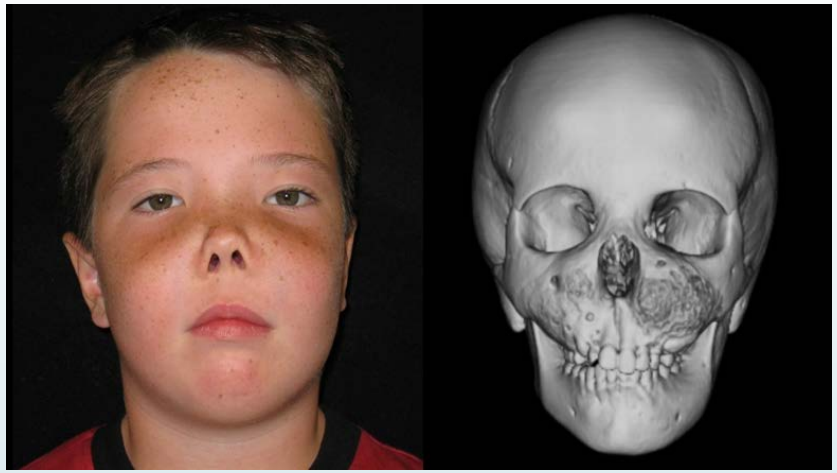
- Decompression surgery:
  - Skull Based=61590, 61595 divided by Infratemporal versus Transtemporal approach
  - Decompression=69720-69725 lateral and/or medial
  - Suture=69740-69745 lateral and/or medial
- Neurorrhaphy=64864-64866 dived by extracranial versus infratemporal
- Cable/Nerve grafts=64885-64886 divided by less or greater than 4cm
- Nerve transposition=64742-facial nerve
- Muscle transfer=15730 midface flap, 15733 myocutaneous or fasciocutaneous head and neck
- Tarsorrhaphy=67875-67882 temporary closure versus contruction of intermarginal adhesions
- Gold weights=67912 Correction of logophtalmos
- Graft for facial nerve paralysis=15840-15845 divided by type of graft

ICD-10-PCS -Congenital Facial Palsy

- 00Q--- Repair, Central Nervous System=Neurorrhaphy
- 00U--- Supplement, Central Nervous System=Cable/Nerve Grafts, Facial Nerve Paralysis
- 00S--- Reposition, Central Nervous System=Nerve Transposition
- 00X---Transfer, Central Nervous System=Nerve Transposition
- 0KX--- Transfer, Muscles
- 08Q--- Repair, Eye=Tarsorrhaphy, Gold Weights

Craniofacial Fibrous Dysplasia-What is it?





- Craniofacial fibrous dysplasia may cause shifting of facial features and facial asymmetry, such as incorrect placement of the eyes, misalignment of the jaw, and other problems.
- There are two forms of fibrous dysplasia:
- Monostotic, which affects one bone and is active while the child is growing but often becomes inactive after puberty
- Polyostotic, which affects multiple bones and may remain active throughout a person's life
- Fibrous dysplasia may appear in childhood, usually between the ages of 3 and 15. Boys are more often diagnosed with fibrous dysplasia than girls, except one specific type of polyostotic fibrous dysplasia — McCune-Albright syndrome — which is more common in girls and affects the bones and skin and is associated with hormonal imbalance and often precocious (premature) puberty.

Craniofacial Fibrous Dysplasia

Diagnoses

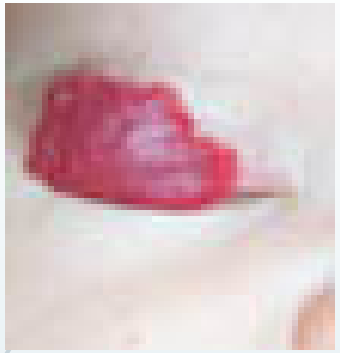
- M27.8= Other specified diseases of jaws
- Q85.5=Other congenital malformation syndromes with other skeletal changes
- M85.00-M85.09=Fibrous dysplasia of bone, Monostotic
- Q87.1=Congenital malformation syndromes predominantly associated with short stature, polyostotic

Procedures

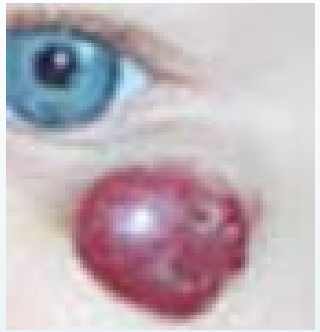
- 21025-21026=Excision of bone divided by mandible or facial bones
- May also contain procedures from 21029-21070 as needed for excision of growths that are not dysplasia such as cysts
- May also contain procedures from 61580-61619 as needed for skull based surgery for procedures on the skull base and the Calveria
- **0NT--- Resection, Head and Facial Bones=Excision of Bone**
- **0NB--- Excision, Head and Facial Bones=Excision of lesions0NC--- Extirpation, Head and Facial Bones= Skull Based Surgery**
- **0NQ--- Repair, Head and Facial Bones=Skull Based Surgery**

Hemangioma-what is it?

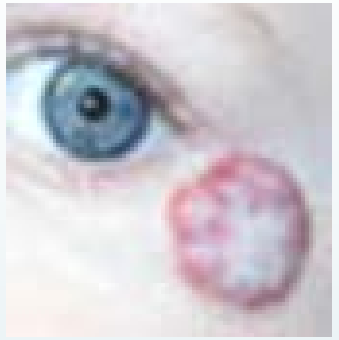
Growth



Resting



Shrinking



- Most hemangiomas are not visible at birth, but they often begin to appear during the first four to six weeks of a child's life. All skin hemangiomas will be visible by six months of age.
- They may occur anywhere on the skin surface, but they are most common on the scalp, face and neck. Many first appear as a small bruise, scratch or a tiny red bump. But unlike other types of birthmarks, they will grow and change quickly during your child's first few months.
- Hemangiomas can occur anywhere on the skin, including, rarely, in the organs of the body. Most often, hemangiomas grow on the skin of the head or neck.
- There are three general types of infantile hemangiomas:
- **Superficial hemangiomas**, which occur on the outer layers of the skin, are typically bright red to purple in color.
- **Deep hemangiomas**, which grow under the skin in the fat, may be blue, purple or even skin color (if they are deep enough under the skin surface).
- **Mixed hemangiomas** are the most common type of hemangioma. These hemangiomas have both superficial and deep components.

Hemangioma

Diagnosis

- D18.01=Hemangioma of skin and subcutaneous tissue
  - Angiolymphoid hyperplasia with eosinophilia
  - Angioma of skin
  - Angioma of subcutaneous tissue
  - Angioma, skin
  - Angioma, subcutaneous tissue
  - Cherry angioma
  - Eyelid hemangioma
  - Hemangioma of eyelid
  - Hemangioma of skin
  - Hemangioma of subcutaneous tissue
  - Senile angioma
  - Strawberry hemangioma
  - Strawberry nevus of skin
- I78.0 Hereditary hemorrhagic telangiectasia
- I78.1 Nevus, non-neoplastic
- Q82.5 Congenital non-neoplastic nevus
- Q85.8 Other phakomatoses, not elsewhere classified
- Q85.9 Phakomatosis, unspecified

Hemangioma

Procedures

- Surgical Excision-based on size and anatomic site
  - 11420-11426 benign lesions of scalp, neck, hands, feet, genitalia
  - 11440-11446 benign lesion of face, ears, eyelids, nose, lips and mucous membranes
- Laser Surgery
  - 17106-17108 divided by size of hemangioma
- **0HB---- Excision, Skin and Breast=Benign Lesion Excision of Skin**
- **0HC--- Extirpation, Skin and Breast=Benign Lesion Excision of Skin**
- **0H0--- Alteration, Skin and Breast=Skin Substitute Tissue Grafts**

Antley-Bixler Syndrome-What is it?

- Antley-Bixler Syndrome is typically characterized by distinctive malformations of the head and facial (craniofacial) area. In most affected infants, there is premature closure of the fibrous joints (sutures) between bones of the front, upper sides, and back portion of the skull (i.e., craniosynostosis involving the coronal and lambdoidal sutures). As a result, the head may appear abnormally short and broad (brachycephalic) and when viewed from above appears like a trapezoid. Additional craniofacial abnormalities may include a large, prominent forehead (frontal bossing, underdeveloped middle regions of the face (midfacial hypoplasia); a large nose with a low nasal bridge; protruding eyes (proptosis); and low-set, malformed (dysplastic) ears.

Antley-Bixler Syndrome-What is it?

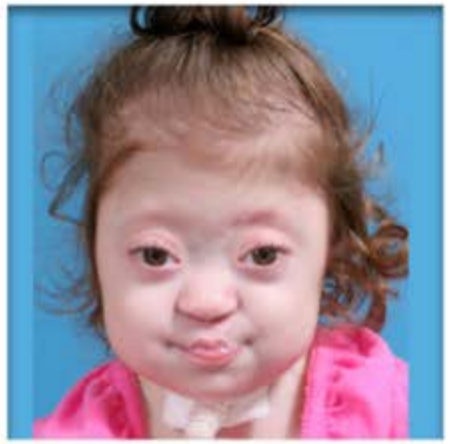
- Antley-Bixler Syndrome is also characterized by additional, distinctive skeletal abnormalities. These may include fusion of adjacent bones of the arms, particularly the forearm bone on the thumb side of the arm (radius) and the long bone of the upper arm (radiohumeral synostosis). In some cases, the forearm bone on the “pinky” side of the arm (ulna) may also be affected. In addition, due to permanent flexion or extension of certain joints in fixed postures (joint contractures), there may be limited movements of the fingers, wrists, ankles, knees, and/or hips. Affected individuals may also have unusually long, thin fingers and toes (digits) with permanent flexion of one or more digits (camptodactyly), malformations of the feet (“rocker-bottom” feet); or bowing and/or fractures of the thigh bones.

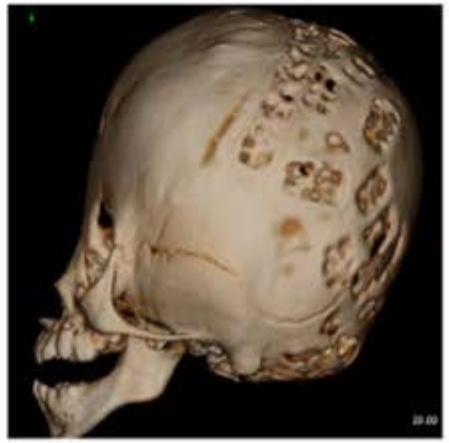
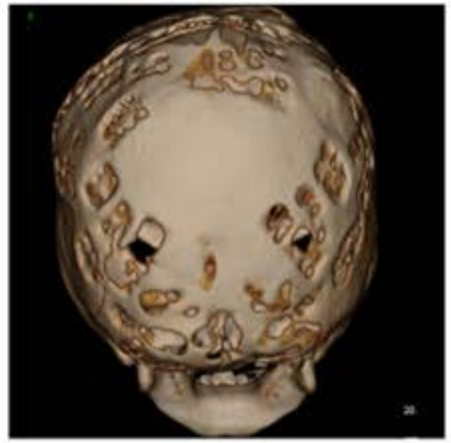
Antley-Bixler Syndrome-What is it?

- In some affected infants, a bony or thin layer of tissue may block the passageway between the nose and throat (choanal stenosis or atresia), leading to difficulties breathing. In such cases, without prompt, appropriate treatment, potentially life-threatening complications may result. There are many affected individuals, however, who live relatively long longs.
- Some individuals with Antley-Bixler Syndrome may have additional physical abnormalities. These may include certain malformations of the heart and/or the urinary and genital organs (urogenital defects).

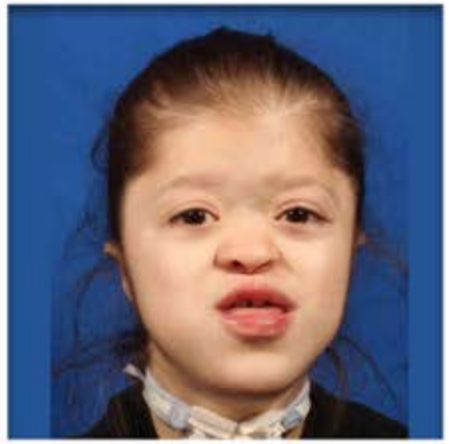
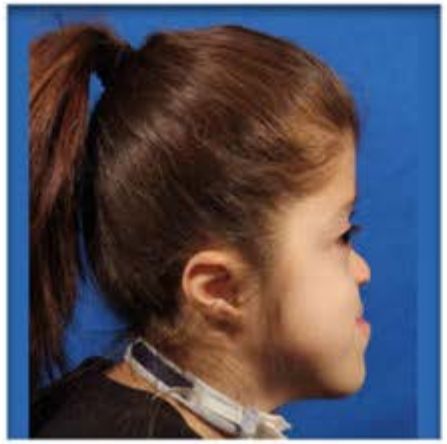
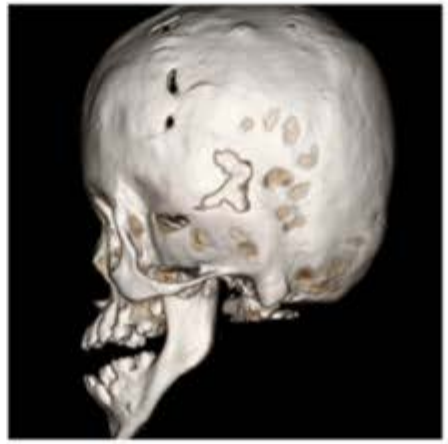
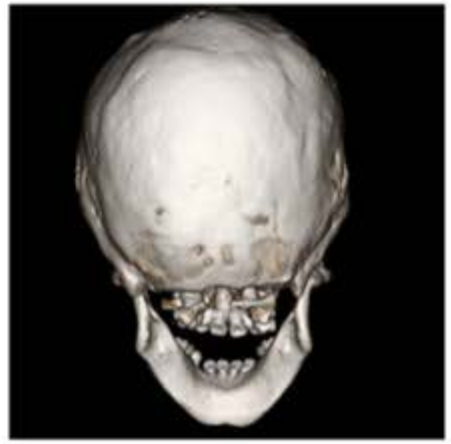
Antley-Bixler Syndrome

Craniofacial
TEAM OF TEXAS



1 year old

8 year old

Antley-Bixler Syndrome

Diagnosis

- Q87.5=Other congenital malformation syndromes with other skeletal changes
  - Other signs and symptoms include:
    - Craniosynostosis=Q75.0
    - Brachycephalic=Q75.0
    - Frontal bossing=Q75.8
    - Midfacial hypoplasia=M26.02
    - Exophthalmos=H05.20
    - Radiohumeral synostosis/ Camptodactyly=Q74.0
    - Joint contractures-dependent of joint affected
    - Choanal stenosis=Q30.0
    - Congenital bowing of femur=Q68.3
    - Congenital bowing of tibia and fibula=Q68.4

Antley-Bixler Syndrome

Procedures

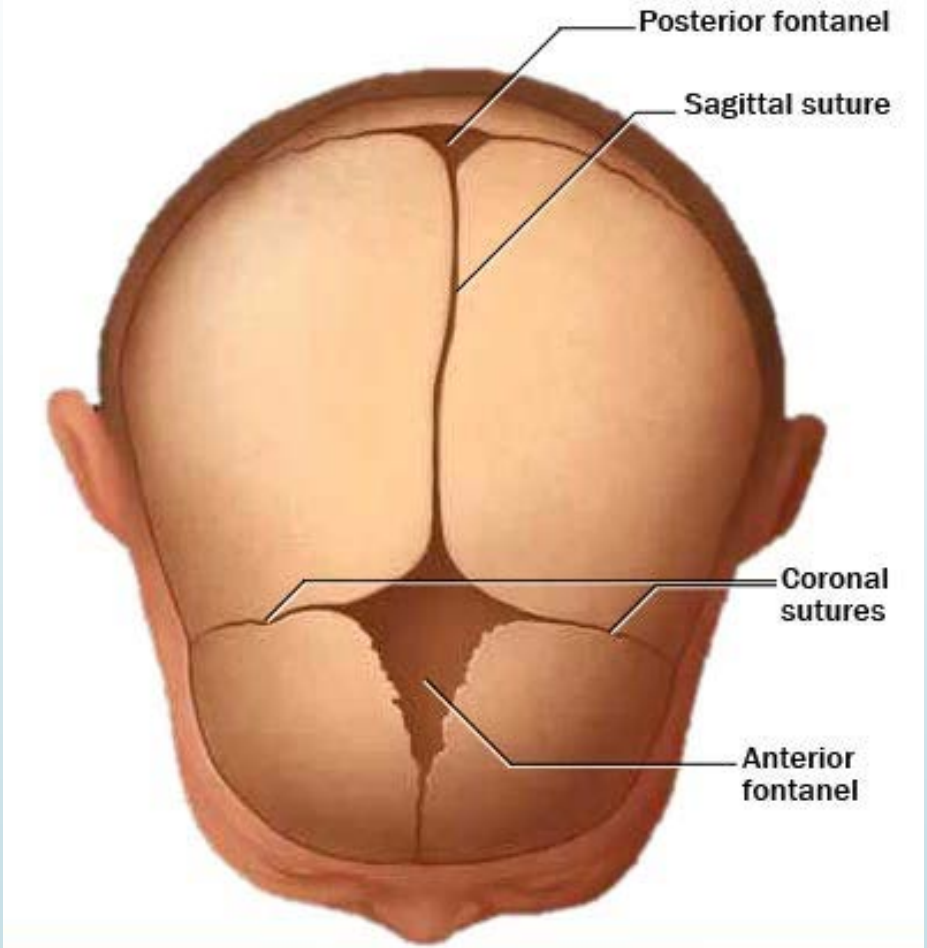
- 30540-30545=Repair choanal atresia divided by intranasal or transpalatine
- 61550-61559=Craniectomy for craniosyntosis divided by anatomic site and number of sutures repaired
- Other surgical procedures as needed for per individual patient
- 96040=Medical genetics and genetic counseling services each 30 minutes….
- 97161-97164=Physical Therapy Evaluations divided by key components
- 97010-97530=Therapeudic modalities and Therapeutic procedures are done as needed per individual patient

ICD-10-PCS - Antley-Bixler Syndrome

- 09Q--- Repair, Ear, Nose, Sinus=Repair of Choanal Atresia
- 0N8--- Division, Head and Facial Bones=Craniectomy/Craniotomy
- F00--- Rehabilitation, Speech Assessment
- F01--- Rehabilitation, Motor and/or Nerve Function Assessment
- F02--- Rehabilitation, Activities of Daily Living Assessment
- F06--- Rehabilitation, Speech Treatment
- F07--- Rehabilitation, Motor Treatment
- F09--- Rehabilitation, Hearing Treatment
- F0B--- Rehabilitation, Cochlear Implant Treatment

Craniosynostosis-What is it?

- Craniosynostosis (kray-nee-o-sin-os-TOE-sis) is a birth defect in which one or more of the fibrous joints between the bones of your baby's skull (cranial sutures) close prematurely (fuse), before your baby's brain is fully formed. Brain growth continues, giving the head a misshapen appearance.
- Craniosynostosis usually involves fusion of a single cranial suture, but can involve more than one of the sutures in your baby's skull (complex craniosynostosis). In rare cases, craniosynostosis is caused by certain genetic syndromes (syndromic craniosynostosis).



Craniosynostosis

Diagnoses

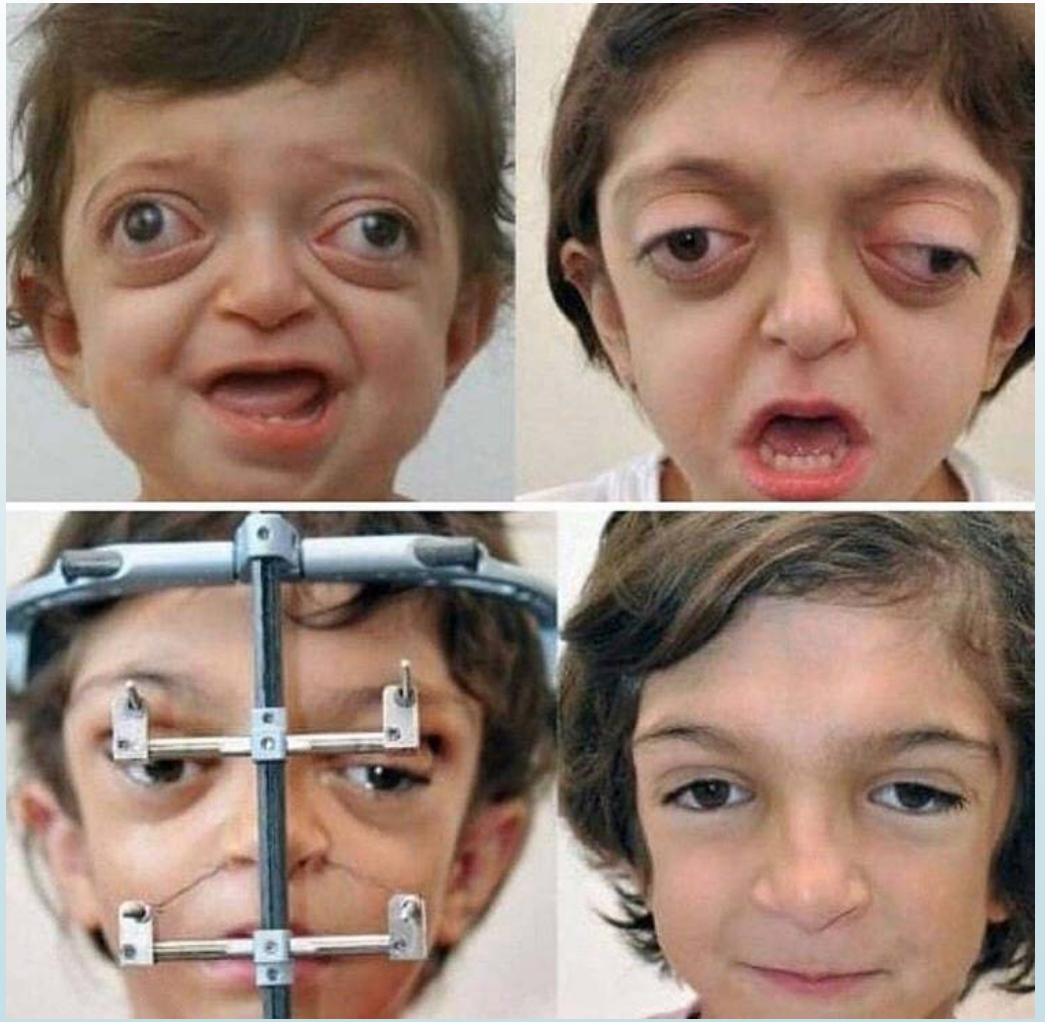
- Craniosynostosis=Q75.0
- Scaphocephaly and Turricephaly=Q75.8

Procedures

- 61550-61559=Craniectomy for craniosyntosis divided by anatomic site and number of sutures repaired
- Other surgical procedures as needed for per individual patient
- 96040=Medical genetics and genetic counseling services each 30 minutes....
- **0NT--- Resection, Head and Facial Bones=Excision of Bone**
- **0NB--- Excision, Head and Facial Bones=Excision of lesions**
- **0NC--- Extirpation, Head and Facial Bones= Skull Based Surgery**
- **0NN---Release, Head and Facial Bones=Craniosynotosis**
- **0NQ--- Repair, Head and Facial Bones=Skull Based Surgery**

Crouzon's Syndrome-What is it?

- **Crouzon syndrome** is a disorder characterized by early fusion of certain skull bones (craniosynostosis). This prevents normal growth of the skull, which can affect the shape of the head and face. Signs and symptoms of Crouzon syndrome may include wide-set, bulging eyes; strabismus (misalignment of the eyes); a small, "beak-shaped" nose; and an underdeveloped upper jaw.[1][2] Other features may include dental problems, hearing loss, and/or cleft lip and palate. The severity of signs and symptoms can vary among affected people, even within a family. Intelligence is usually normal, but intellectual disability may be present.[1][3] Crouzon syndrome is caused by changes (mutations) in the *FGFR2* gene and is inherited in an autosomal dominant manner. Treatment may involve surgeries to prevent complications, improve function, and aid in healthy psychosocial development.



Crouzon's Syndrome

Diagnoses

- Craniosynostosis=Q75.0
- Scaphocephaly and Turricephaly=Q75.8
- Cleft lip and/or Palate=Q35.1-Q35.9
- Proptosis=H05.20
- Primary sleep apnea of newborn=P28.3
- Sleep apnea=G47.3
- Other Congenital Hydrocephalus=Q03.8
- Congenital Hydrocephalus=Q03.9
- Strabismus=H50.00-H50.9

Procedures

- 61550-61559=Craniectomy for craniosyntosis divided by anatomic site and number of sutures repaired
- Extraocular Muscle Surgery for Strabismus 67311-67346
- Procedures done for Cleft lip and or Cleft Palate

Crouzon's Syndrome

Procedures

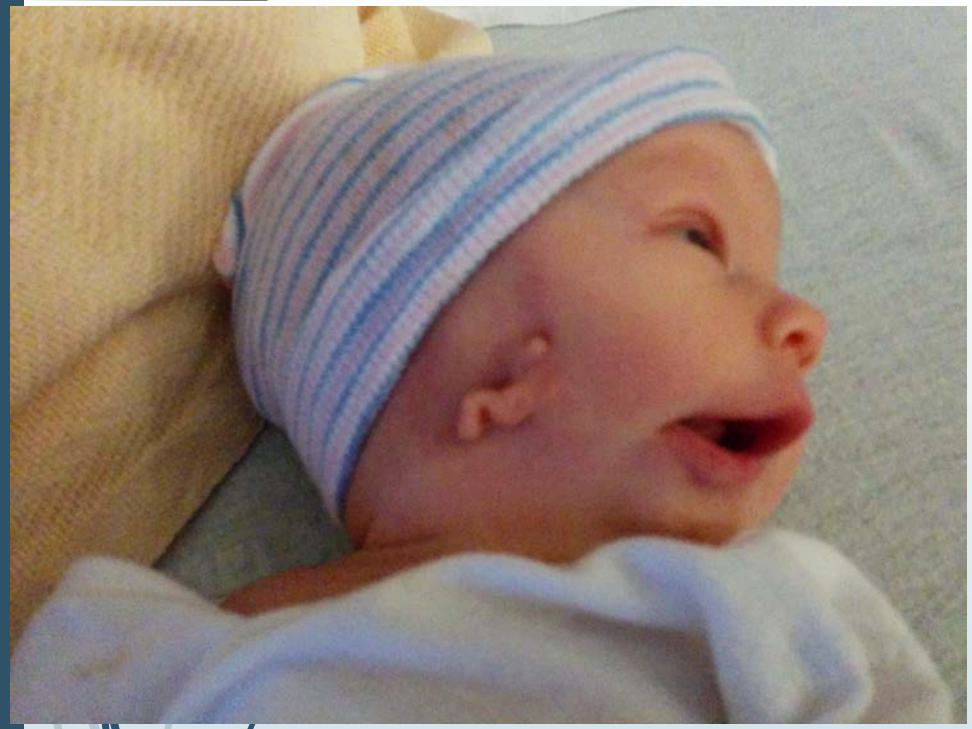
- 20690 Application of a uniplane (pins or wires in 1 plane), unilateral, external fixation system
- 20692 Application of multiplane (pins or wires in more than one plane), unilateral, external fixation system (e.g., Ilizarov, Monticelli type)
- 20693 Adjustment of revision of external fixation system requiring anesthesia (eg, new pin[s] or wire[s] and/or new ring[s] or bar[s]
- 20694 Removal, under anesthesia, of external fixation system
- 20696 Application of multiplane (pins or wires in more than 1 plane), unilateral, external fixation with stereotactic computer-assisted adjustment (eg, spatial frame), including imaging; initial and subsequent alignment(s), assessment(s), and computation(s) of adjustment schedule(s)
- 21100 Application of halo type appliance for maxillofacial fixation, includes removal (separate procedure)
- 21110 Application of interdental fixation device for conditions other than fracture or dislocation, includes removal
- 21195 Reconstruction of mandibular rami and/or body, sagittal split; without internal rigid fixation

ICD-10-PCS - Crouzon's Syndrome

Procedures

- **0WH--- Insertion, Anatomical Regions, General=Application of Fixation Device without Fracture**
- **0NS--- Reposition, Head and Facial Bones=Application of Fixation Device with Fracture**
- **0NW--- Revision, Head and Facial Bones=Revision of External Fixation**
- **0NP--- Removal, Head and Facial Bones=Removal External Fixation**
- **0NQ--- Repair, Head and Facial Bones=Reconstruction of Mandibular Rami**
- **0NN---Release, Head and Facial Bones=Craniosyntosis**
- **08Q--- Repair, Eye=Strabismus Surgery**

Goldenhar Syndrome/Hemifacial microsomia-What is it?



- **Goldenhar disease** is a condition that is present at birth and mainly affects the development of the eye, ear, and spine. The main sign and symptoms are facial asymmetry (one side of the face is different from the other), a partially formed ear (microtia) or totally absent ear (anotia), noncancerous (benign) growths of the eye (ocular dermoid cysts), and spinal abnormalities. Goldenhar disease may also affect the heart, lungs, kidneys, and central nervous system.[1][2] It is due to problems that occur when the fetus is forming within the womb of the mother, in structures known as the first and second brachial arch. These structures will develop to form the neck and the head. The cause is still unknown.[1][2][3][4] Goldenhar syndrome is part of a group of conditions known as craniofacial microsomia. It is not known whether the conditions included in the group really are different conditions or part of the same problem with different degrees of severity. Treatment is age-dependent, with interventions at appropriate stages during the growth and development of the skull and face

Goldenhar Syndrome/Hemifacial microsomia

Diagnoses

- Other specified congenital malformations of skull and face bones=Q75.8
- Q67.0=Congenital facial asymmetry
- Code any associated manifestations such as:
- Microtia=Q17.2
- Auditory Atresia=Q16.1
- Cleft Lip and/or Palate=Q35.1-Q35.9
- Eipbulbar tumors:
  - **H21.32 Implantation cysts of iris, ciliary body or anterior chamber**
  - H21.321 ..... right eye
  - H21.322 ..... left eye
  - H21.323 ..... bilateral
  - H21.329 ..... unspecified eye
- Macrostomia-Q18.4
- Microstomia=Q18.5
- Hearing Loss=H90.0-H90.A32
  - https://www.icd10data.com/ICD10CM/Codes/H60-H95/H90-H94/H90-
- Tongue Tie= Q38.1
- Other complications include:
  - Dental abnormalities
  - Spinal and Limb Malformations
  - Heart, Kidney and Lung problems
  - Hydrocephalus with or without Intellectual Disability

Goldenhar Syndrome/Hemifacial microsomia

Procedures

- 14040 Adjacent tissue transfer or rearrangement, forehead, cheeks, chin mouth, neck, axillae, genitalia
- hands and/or feet; defect 10sq cm or less
- 14041 defect 10.1 sq cm to 30.0 sq cm.
- 14060 Adjacent tissue transfer or rearrangement, eyelids, nose, ears, and/or lips; defect 10 sq cm or
- less
- 14061 defect 10.1 sq cm to 30.0 sq cm
- 15120 - 15261 (additional reconstructive codes under the Integumentary System)
- 15576 Formation of direct or tubed pedicle, with or without transfer; eyelids, nose, ears, lips, or intraoral
- 20902 Bone harvest any area, major or large.
- 41115 Excision of lingual frenum (frenectomy)

Goldenhar Syndrome/Hemifacial microsomia

Procedures

- 20690 Application of a uniplane (pins or wires in 1 plane), unilateral, external fixation system
- 20692 Application of multiplane (pins or wires in more than one plane), unilateral, external fixation system (e.g., Ilizarov, Monticelli type)
- 20693 Adjustment of revision of external fixation system requiring anesthesia (eg, new pin[s] or wire[s] and/or new ring[s] or bar[s]
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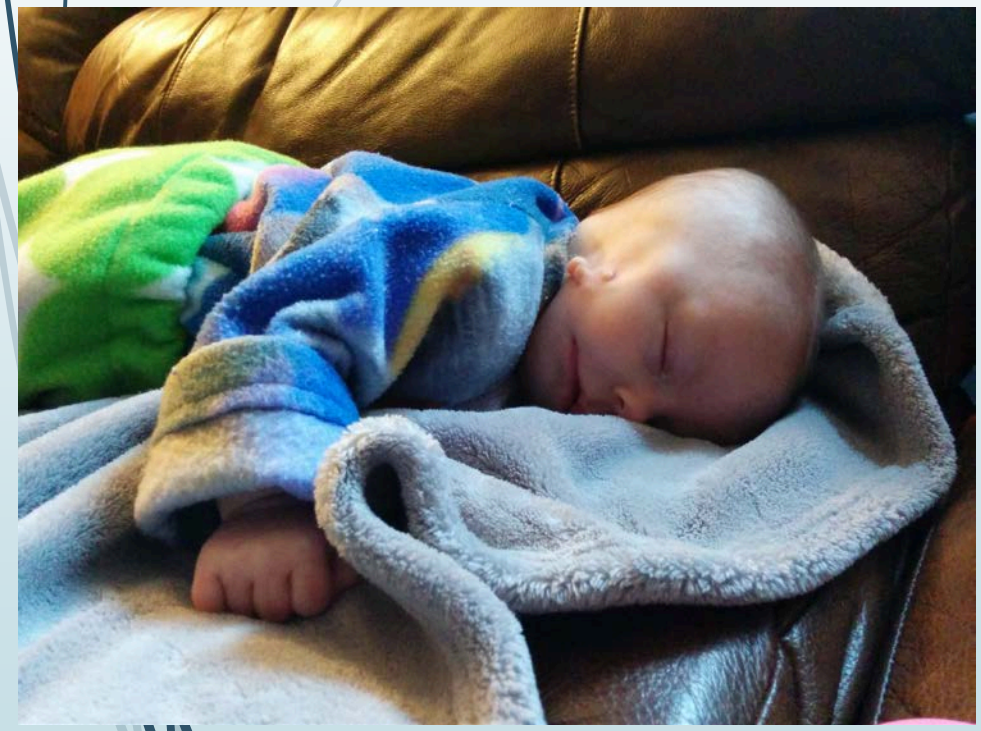
- 20696 Application of multiplane (pins or wires in more than 1 plane), unilateral, external fixation with stereotactic computer-assisted adjustment (eg, spatial frame), including imaging; initial and subsequent alignment(s), assessment(s), and computation(s) of adjustment schedule(s)
- 21100 Application of halo type appliance for maxillofacial fixation, includes removal (separate procedure)
- 21110 Application of interdental fixation device for conditions other than fracture or dislocation, includes removal
- 21195 Reconstruction of mandibular rami and/or body, sagittal split; without internal rigid fixation

Goldenhar Syndrome/Hemifacial microsomia



Procedures

- 92585 Auditory evoked potentials for evoked response audiometry and/or testing of the central nervous system; comprehensive
- 92586 limited
- 92507 Treatment of speech, language, voice, communication, and/or auditory processing disorder; individual
- 69310 Reconstruction of external auditory canal (meatoplasty) (eg, for stenosis due to injury, infection) (separate procedure)
- 69320 Reconstruction external auditory canal for congenital atresia, single stage

ICD-10-PCS-Goldenhar Syndrome/Hemifacial microsomia

- 0JX---- Transfer, Subcutaneous Tissue and Fascia
- 0HRX-- Replacement, Skin and Breast
- 0H0--- Alteration, Skin and Breast=Skin Substitute Grafts
- 0HW--- Revision, Skin and Breast=Revision of Skin Grafts
- 0HX--- Transfer, Skin and Breast=Autologous Skin Grafts
- 0QS---- Reposition, Lower Bones=Bone graft
- 0PS---- Reposition, Upper Bones=Bone graft
- 0NU---- Reposition, Head and Facial Bones=Bone graft
- 0H0--- Alteration, Skin and Breast=Skin Substitute Tissue Grafts
- 0CN7– Release, Mouth and Throat=Frenectomy

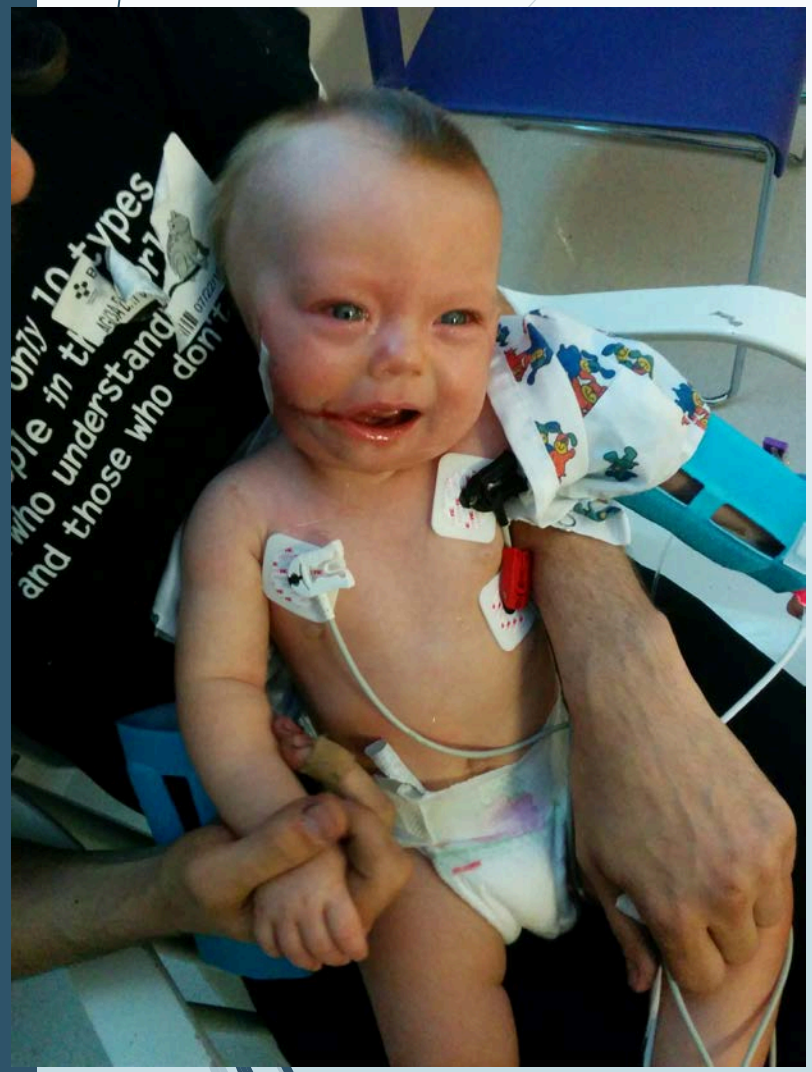
ICD-10-PCS-Goldenhar Syndrome/Hemifacial microsomia

- 0CDWXZ- Extraction, Upper Tooth
- 0CDXXZ- Extraction, Lower Tooth
- 0WH--- Insertion, Anatomical Regions, General=Application of Fixation Device without Fracture
- 0NS--- Reposition, Head and Facial Bones=Application of Fixation Device with Fracture
- 0NW--- Revision, Head and Facial Bones=Revision of External Fixation
- 0NP--- Removal, Head and Facial Bones=Removal External Fixation
- 0NQ--- Repair, Head and Facial Bones=Reconstruction of Mandibular Rami
- 0H8--- Division, Skin and Breast=Direct or Tubed Pedicle flap
- 0NR--- Replacement, Head and Facial Bones=Bone Harvesting
- 09Q--- Repair, Ear, Nose, Sinus=Reconstruction of External Auditory Canal

ICD-10-PCSGoldenhar Syndrome/Hemifacial microsomia

- 0B1--- Bypass, Respiratory System=Tracheostomy
- 0B2--- Change, Respiratory System=Change of Tracheostomy
- 3E00-- Tattooing/Intradermal Pigments
- F00--- Rehabilitation, Speech Assessment
- F06--- Rehabilitation, Speech Treatment
- F09--- Rehabilitation, Hearing Treatment
- F0B--- Rehabilitation, Cochlear Implant Treatment
- F0D--- Rehabilitation, Device Fitting
- F13---Diagnostic Audiology, Hearing Assessment
- F14--- Diagnostic Audiology, Hearing Aid Assessment

Goldenhar Syndrome/Hemifacial macrosomia



Treacher-Collins Syndrome-What is it?

- **Treacher Collins syndrome (TCS)** is a condition that affects the development of bones and other tissues of the face. The signs and symptoms vary greatly, ranging from almost unnoticeable to severe. Most affected people have underdeveloped facial bones, particularly the cheek bones, and a very small jaw and chin (micrognathia). Other features may include cleft palate, eye abnormalities, and hearing loss.[1] TCS may be caused by mutations in the *TCOF1*, *POLR1C*, or *POLR1D* genes.[2] When the *TCOF1* or *POLR1D* gene is responsible, it is inherited in an autosomal dominant manner.[2] However, about 60% of autosomal dominant cases are due to a new mutation in the gene and are not inherited from a parent.[1] When the *POLR1C* gene is responsible, it is inherited in an autosomal recessive manner.[2] In some cases, the genetic cause of the condition is unknown

Treacher-Collins Syndrome-What is it?

- The signs and symptoms of Treacher Collins syndrome vary greatly, ranging from almost unnoticeable to severe. Most affected people have underdeveloped facial bones, particularly the cheek bones, and a very small jaw and chin (micrognathia). Some people with this condition are also born with an opening in the roof of the mouth called a cleft palate. In severe cases, underdevelopment of the facial bones may restrict an affected infant's airway, causing potentially life-threatening respiratory problems.[3]People with Treacher Collins syndrome often have eyes that slant downward, sparse eyelashes, and a notch in the lower eyelids called a coloboma. Some people have additional eye abnormalities that can lead to vision loss. The condition is also characterized by absent, small, or unusually formed ears. Defects in the middle ear (which contains three small bones that transmit sound) cause hearing loss in about half of affected people.

Treacher-Collins Syndrome

Diagnoses

- Mandibulofacial dysostosis=Q75.4
- Hearing Loss=H90.0-H90.A32
  - https://www.icd10data.com/ICD10CM/Codes/H60-H95/H90-H94/H90-
- Cleft Lip and/or Palate=Q35.1-Q35.9
- Coloboma Eyelid=Q10.3
- Coloboma Iris=Q13.0
- Auditory Atresia=Q16.1
- Cleft Lip and/or Palate=Q35.1-Q35.9
- Strabismus=H50.00-H50.9

Treacher-Collins Procedures

- 14040 Adjacent tissue transfer or rearrangement, forehead, cheeks, chin mouth, neck, axillae, genitalia,
- hands and/or feet; defect 10sq cm or less
- 14041 defect 10.1 sq cm to 30.0 sq cm.
- 14060 Adjacent tissue transfer or rearrangement, eyelids, nose, ears, and/or lips; defect 10 sq cm or
- less
- 14061 defect 10.1 sq cm to 30.0 sq cm
- 15120 - 15261 (additional reconstructive codes under the Integumentary System)
- 15576 Formation of direct or tubed pedicle, with or without transfer; eyelids, nose, ears, lips, or intraoral
- 20902 Bone harvest any area, major or large.
- 69310 Reconstruction of external auditory canal (meatoplasty) (eg, for stenosis due to injury, infection) (separate procedure)
- 69320 Reconstruction external auditory canal for congenital atresia, single stage

Treacher-Collins Procedures

- 20690 Application of a uniplane (pins or wires in 1 plane), unilateral, external fixation system
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- 21100 Application of halo type appliance for maxillofacial fixation, includes removal (separate procedure)
- 21110 Application of interdental fixation device for conditions other than fracture or dislocation, includes removal
- 21195 Reconstruction of mandibular rami and/or body, sagittal split; without internal rigid fixation
- 31600 Tracheostomy, planned (separate procedure);
- 36101 younger than 2 years

ICD-10-PCS-Treacher-Colllins

- 0JX---- Transfer, Subcutaneous Tissue and Fascia
- 0HRX-- Replacement, Skin and Breast
- 0H0--- Alteration, Skin and Breast=Skin Substitute Grafts
- 0HW--- Revision, Skin and Breast=Revision of Skin Grafts
- 0HX--- Transfer, Skin and Breast=Autologous Skin Grafts
- 0QS---- Reposition, Lower Bones=Bone graft
- 0PS---- Reposition, Upper Bones=Bone graft
- 0NU---- Reposition, Head and Facial Bones=Bone graft
- 0H0--- Alteration, Skin and Breast=Skin Substitute Tissue Grafts
- 0CN7– Release, Mouth and Throat=Frenectomy

ICD-10-PCS-Teacher-Collins

- 0CDWXZ- Extraction, Upper Tooth
- 0CDXXZ- Extraction, Lower Tooth
- 0WH--- Insertion, Anatomical Regions, General=Application of Fixation Device without Fracture
- 0NS--- Reposition, Head and Facial Bones=Application of Fixation Device with Fracture
- 0NW--- Revision, Head and Facial Bones=Revision of External Fixation
- 0NP--- Removal, Head and Facial Bones=Removal External Fixation
- 0NQ--- Repair, Head and Facial Bones=Reconstruction of Mandibular Rami
- 0H8--- Division, Skin and Breast=Direct or Tubed Pedicle flap
- 0NR--- Replacement, Head and Facial Bones=Bone Harvesting
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ICD-10-PCS-Treacher-Collins

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- F09--- Rehabilitation, Hearing Treatment
- F0B--- Rehabilitation, Cochlear Implant Treatment
- F0D--- Rehabilitation, Device Fitting
- F13---Diagnostic Audiology, Hearing Assessment
- F14--- Diagnostic Audiology, Hearing Aid Assessment

Connecticut Children's Medical Center Craniofacial Team

- Craniofacial Team: https://www.connecticutchildrens.org/search-specialties/craniofacial-program/craniofacial-program-our-team/
- Services: https://www.connecticutchildrens.org/search-specialties/craniofacial-program/craniofacial-program-services/
- Resources: https://www.connecticutchildrens.org/search-specialties/craniofacial-program/craniofacial-program-resources/
- Beyond the Face Documentary- feature CCMC Craniofacial patients https://www.connecticutchildrens.org/search-specialties/craniofacial-program/craniofacial-beyond-the-face/

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- https://www.aaoms.org/images/uploads/pdfs/cleft_lip_and_palate.pdf
- http://www.stlouischildrens.org/our-services/plastic-surgery/photo-gallery/cleft-nasal-reconstruction-gallery
- https://emedicine.medscape.com/article/878464-overview#a7
- https://www.chop.edu/conditions-diseases/craniofacial-fibrous-dysplasia
- https://www.healthline.com/health/hemangioma-of-skin#treatment
- https://rarediseases.org/rare-diseases/antley-bixler-syndrome/
- http://craniofacialteamtexas.com/craniofacial-gallery/craniofacial-syndromes-gallery/
- https://www.mayoclinic.org/diseases-conditions/craniosynostosis/symptoms-causes/syc-20354513
- https://rarediseases.info.nih.gov/diseases/6206/crouzon-syndrome
- https://www.chop.edu/conditions-diseases/crouzon-syndrome
- https://www.supercoder.com/webroot/upload/general_pages_docs/document/mm_0407_coveragepositioncriteria_craniofacial_remodeling_with_do.pdf
- https://rarediseases.info.nih.gov/diseases/9124/treacher-collins-syndrome
- https://www.adelaidenow.com.au/business/sa-business-journal/worlds-most-famous-treacher-collins-sufferer-inspires-zack-2-of-mannum/news-story/5a7737ac9d0591122a8448b087567c65
- https://www.facebook.com/Jono-Lancaster-179404148779245/

Extra Resources for Further Knowledge

- Special Books by Special Kids-A YouTube Channel for different and rare diseases, disorders and syndromes: https://www.youtube.com/channel/UC4E98HDsPXrf5kTKIgrSmtQ
- Children's Craniofacial Association: https://ccakids.org/
- World Craniofacial Organization: https://worldcf.org/
- FACES National Craniofacial Association: http://www.faces-cranio.org/
- American Cleft Palate-Craniofacial Association: https://acpa-cpf.org/
- My Face: https://www.myface.org/
- American Society of Craniofacial Surgeons: http://ascfs.org/