A classification of cranio facio cervical (branchial) clefts (Bangalore classification)

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ABSTRACT

This new classification is based on the analysis of the following:
1. The study of Embryological developments of the normal structures and the clefts in the head and neck regions,
2. The Clinical presentation of Clefts in the head and neck regions in our series of 146 cases,
3. The study of clefts under Rare craniofacial, Branchial [Cervical] and Classifications by various Authors and
4. The review of Literature pertaining to the clefts in head and neck regions.

The documentation of commonly diagnosed and treated cleft lip and palate anomalies have remained unsatisfactory. As the regular cleft lip and palate falls within the purview of this new classification, a separate classification of Rare Craniofacial clefts can be avoided. This is an attempt made to bring all varieties of cleft deformities in the head and neck region under one Classification and to plan and execute better techniques in the field of assessment and management.

KEY WORDS

Schisis, Craniorachischisis, Rhinoschisis, Meloschisis, Blepharoschisis, Glossoschisis

INTRODUCTION

The incidence of clefts in the head and neck region is not rare. The cleft may occur in isolation or in combination with soft and skeletal tissues. The clefts may be seen in variable degrees on either side [unilateral] of the midline or on both sides [bilateral]. Some of the clinically diagnosed and treated cases of cleft lip and palate, have shown other type of hidden clefts in the face on further investigations at the time of pre/post operative follow up after this study was taken up.

Varieties of separate classifications are projected with regard to cleft lip and palate. The clefts falling under the category of "Rare Craniofacial Clefts" are also classified by many authors differently. One of the popular classifications is that projected by Sir Paul Tessier's as "Rare Cranio Facial Clefts". The central point of reference in this classification is the orbit.

Keeping in view the fact that most of the clefts in our series are radiating from the oral cavity outwards, the oral cavity is taken as the central point of reference for the Anterior...
Median and Oblique Craniofacial, Cervical [Branchial], Lower Median Facio Mandibular and Median Cervical Clefts. Those Transverse clefts that occur in the region of the orbit [IX and X] and the Posterior clefts in the head and neck region [IA and IB] have no relation to the oral cavity as the point of reference. To simplify, the Numbers from I to XV are used to denote the clefts in the soft tissue and skeletal structures in the head and neck regions. The clefts are required to be described as left sided [L] or right sided [R] or bilateral [L and R]. The clefts in the left side are numbered in clockwise direction and the clefts in the right are numbered in the anti clockwise direction from midline outwards. The clefts may be isolated soft tissue clefts [S] or skeletal [Sk] or combined clefts [S and Sk]. The clefts may be in varying degrees as incomplete or complete in soft and skeletal tissues or combined [Table 1 and 2, and Figures 1, 2 & 3].

**MATERIALS AND METHODS**

This new Classification is arrived at on the basis of analysis of the following:


<table>
<thead>
<tr>
<th>Table 1: Classification of cranio facio cervical clefts</th>
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<td><strong>1. Soft tissue</strong></td>
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<td><strong>A. ANTERIOR</strong></td>
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<td><strong>Group</strong></td>
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<td>1. Upper Median - Head</td>
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<td>2. Lower Median - Cervical</td>
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2. The study of investigations: Radiological in 126 cases, CT Scan in 46 cases, 3D Scan in 24 cases, MRI in one case, Genetic Karyotype studies in 7 cases and Echo Cardiogram in 4 cases.

3. The study of presentation of various Cranio Facial and Cervical Clefts described under 'Rare Craniofacial Clefts, Branchial Cleft Anomalies and other classifications and

4. A review of clinical cases and literature pertaining to the various clefts including those in the Posterior aspects of head and neck region.

**EMBRYOLOGY**

The head and neck region development occurs during 4th and 8th weeks of embryonic life. The facial primordia begin to appear early in the fourth week around the large stomodeum. The face is formed by the fusion of five swellings [primordia], i.e., an unpaired frontonasal process, a pair of maxillary processes and a pair of mandibular processes. The Pharyngeal Apparatus contributes extensively to the formation of the face, nasal cavities, mouth, larynx, pharynx and neck. Much of the human face and neck is derived from the ancient gill apparatus. Most...
congenital anomalies in this region originate during transformation of the pharyngeal apparatus into its adult derivatives.

During the fifth week, the second pharyngeal arch enlarges and overgrows the third and fourth arches, forming an ectodermal depression - the cervical sinus. The pharyngeal grooves disappear except for the first pair, which persist as the external acoustic meatus. The pharyngeal membranes also disappear, except for the first pair, which become the tympanic membranes. The Branchial anomalies result from persistence of parts of the pharyngeal apparatus that normally disappear. The Branchial cleft anomalies include branchiogenic sinuses, cartilaginous rests, fistulae and cervical cysts. There are four Branchial cleft anomalies described. The First Branchial cleft anomalies are above the level of hyoid and the external orifice is near the auricle or beneath the mandibular ramus. The Second Branchial cleft anomalies are the most common. The external opening is near the junction of the middle and lower thirds of the sternocleidomastoid muscle. The Third branchial cleft anomalies are rare. The external orifice may be located in a similar manner as that of the Second Branchial cleft fistulae, along the anterior border of the lower half of the sternocleidomastoid muscle. The Fourth Branchial cleft anomalies are rare. The internal openings of all the above four Branchial Cleft anomalies open in the tonsillar fossa and piriform sinus.

The thyroid gland develops from the thyroglossal duct,
extending from the foramen cecum in the posterior midline of the tongue through the hyoid bone to the midline of the lower neck. The Thyroglossal duct cysts and sinuses may form anywhere along the course followed by the thyroglossal duct during descent of the thyroid gland from the tongue. Normally the thyroglossal duct atrophies and disappears, but remnants of it may persist and form a cyst in the tongue or in the anterior part of the neck, usually below the hyoid bone. The thyroglossal sinus opens in the median plane of the neck, usually below the hyoid bone. The thyroglossal sinus opens in the median plane of the neck. Incomplete fusion of the distal tongue buds results in a deep median sulcus or cleft in the tongue [glossoschisis].

The primordial mouth or stomodeum initially appears as a slight depression of the surface ectoderm. The development of five facial primordial are focused towards the stomodium. Clefts in the head and neck region occur during this period as a result of non fusion of facial primordial swellings/processes. The factors identified as the probable causes of cleft formation are broadly divided into (1) Genetic factors and (2) Environmental factors. Many common congenital anomalies are caused by genetic and environmental factors acting together - multifactorial inheritance.

The four Branchial [Cervical] Cleft anomalies in the anterior aspect and the clefts described as ‘Cranorachischisis’ on the posterior aspect of the head [cranioschisis] and neck [rachischisis] region occur during the same period of intrauterine life also deserve inclusion in this broad based new classification.
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CLASSIFICATION

"An Embryological and Clinical Classification of Cranio Facio Cervical [Branchial] Clefts"

The Description of Clefts in the Anterior aspect of head and neck.

Number I

Number I is the upper median facio cranial cleft. The soft tissue cleft starts from the oral cavity, runs in an upward and median plane involving the upper lip, nose [Rhinorschisis], naso frontal [Anterior sincipital encephalocele in Figure 4C] and frontal regions [Table 1 and in Figures 1, 2 and 4]. The cleft may involve all of the above soft tissue structures [S] as an isolated cleft or may involve all of the above tissue structures. The Skeletal Cleft [Sk] starts from the oral cavity, runs through the maxilla, nasal, ethmoid, vomer and frontal bones in the midline [Table 2 and in Figures 3 and 4]. The cleft may involve one of the above cranial bones as isolated cranial bony cleft or may involve all the above cranial bones. The cleft may occur as an isolated soft tissue or as an isolated skeletal or in combination of both structures.

Number II

Number II is the Oblique facio cranial cleft. The soft tissue cleft runs in the upward and oblique direction from the oral cavity, involving the medial one third of the upper lip, nose [Rhinorschisis] and forehead [Table 1 and in Figures 1, 2 and 5]. The cleft may involve one of the above soft tissue structure [S] as an isolated cleft or may involve all of the above soft tissue structures. The Skeletal cleft [Sk] starts from the oral cavity runs in upwards and oblique direction,
Number III

Number III is the Oblique facio cranial cleft. The soft tissue cleft \([S]\) runs in the upward and oblique direction from the oral cavity, involving the medial one third of upper lip \([\text{lateral to Number II}]\), in the junction of middle and lateral third of nose \([\text{Rhinosophis}]\), medial third of lower and upper eyelids \([\text{Blepharosophis}]\) and in the medial third of forehead \([\text{Table 1 and in Figures 1, 2 and 6}]\). The cleft may involve one of the above soft tissues as an isolated cleft or may involve all of the above soft tissue structures. The skeletal cleft \([\text{Sk}]\) starts from the oral cavity runs in an upward and oblique direction, involving maxilla, lateral aspect of nasal, lacrimal, palatine, sphenoid and frontal bones \([\text{Table 2 and in Figures 3 and 6}]\). The cleft may involve one of the above cranial bones as an isolated cleft or may involve all the above cranial bones. The cleft may occur as an isolated soft tissue or skeletal cleft or as a combination of both structures. The cleft may be unilateral or bilateral.

Number IV

Number IV is the oblique facio cranial cleft. The soft tissue cleft \([S]\) runs in the upward and oblique direction from the oral cavity, involving the junction of the medial and middle third of the upper lip, face \([\text{Melosophis}]\) at alar groove, middle third of the lower and upper eyelids.
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a partial cleft or may involve all of the above soft tissue structures. The skeletal cleft [Sk] starts from the oral cavity runs in an upwards and oblique direction, involving the lateral aspect of maxilla, sphenoid, zygoma and frontal [middle third] bones [Table 2 and in Figures 3 and 7]. The cleft may involve one of above cranial bones as isolated cleft or involve all bones or in combination of both soft tissue and skeletal structures. The cleft may be unilateral or bilateral.

**Number V**

Number V is the oblique facio cranial cleft. The soft tissue cleft [S] runs in the upward and oblique direction from the oral cavity, involving the upper lip at the junction of middle and lateral third, face [Meloschisis] lateral to the nasolabial groove, middle third of lower and upper eyelids [Blepharochisis or Colomba eyelids] and in the middle third of forehead [Table 1 and in Figures 1, 2 and 8]. The cleft may involve one of the above soft tissue structures as a partial cleft or may involve all of the above soft tissue structures as complete cleft. The skeletal cleft [Sk] starts from the oral cavity, runs in upward and oblique direction,
involving lateral aspect of maxilla, sphenoid, zygoma and frontal and parietal bones [Table 2 and in Figures 3 and 8]. The cleft may involve one of the above cranial bones as a partial cleft or all of the bones or in combination of both soft tissue and skeletal structures. The cleft may be unilateral or bilateral.

**Number VI**

Number VI is the oblique facio cranial cleft. The soft tissue cleft [S] runs in the upward and oblique direction, involving the lateral third of the upper lip, face [Meloschisis], lateral third of lower and upper eyelids [Blepharoschisis] and in the lateral third of the fronto parietal regions [Table 1 and in Figures 1, 2 and 9]. The cleft may involve one of the above structures as a partial cleft or may involve all of the above soft tissue structures as a complete cleft. The skeletal cleft [Sk] starts from the oral cavity runs in an upward and oblique direction, involving the zygoma, temporal and parietal bones [Table 2 and in Figures 3 and 9]. The cleft may involve one of the above cranial bones as a partial or complete cleft of all bones or in combination with both soft tissue and skeletal structures. The cleft may be unilateral or bilateral.

**Number VII**

Number VII is the oblique facio cranial cleft. The soft tissue cleft [S] runs in the upward and oblique direction, involving the lateral third of upper lip, face [Meloschisis], lateral to the lateral canthal region, and temporo parietal region [Table 1 and in Figures 1, 2 and 10]. The cleft may involve one of the above structures as an isolated cleft or may involve all of the above soft tissue structures. The skeletal cleft [Sk] starts from the oral cavity runs in an upward and oblique direction, involving the zygoma, temporal and parietal bones [Table 2 and in Figures 3 and 10]. The cleft may involve one of the above cranial bones or in combination with both soft tissue and skeletal structures, the cleft may be unilateral or bilateral.

**Number VIII**

Number VIII is the oblique facio cranial cleft. The soft tissue

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cleft [S] runs in an upward and oblique direction, involving commissure, face [Meloschisis] and external ear [auricle] [Table 1 and in Figures 1, 2 and 11]. The cleft may extend from the commissure to the external ear cartilages. The cleft in the upper three fourths of the ear occurs in soft tissue and auricular cartilages. In the middle, it may pass through the external auditory canal involving both soft tissue and cartilage. On the lower level it may pass through soft tissue only i.e. lobule. The cleft may involve one of the above soft tissues as an isolated cleft or all the above structures. The cleft in the upper three fourth of the External ear occurs together in soft tissue and skeletal [Auricular cartilage] structures [S-Sk]. The external auditory canal cleft involves both soft tissues and cartilage, and the cleft in the lower one fourth of the external ear is the only soft tissue ear lobe cleft. The skeletal cleft [Sk] runs in upward and oblique direction, involving ramus of the mandible [Table 2 and in Figures 3 and 11]. The cleft may occur as an isolated cleft in the commissure and face as macrostomia, or may involve in a combination of commissure, face and external ear. The cleft may occur one side or on both sides. Clinically unilateral or bilateral cleft through commissure may be seen as macrostomia.

Number IX

Number IX is the Transverse medial ocular facio cranial cleft. The soft tissue cleft [S] runs in the transverse direction from the medial canthal region towards the median plane,

Figure 13: No. X.- Cranio Facio Cervical (Branchial) Clefts – 1.Soft tissue A. Anterior, Group – 3. Transverse facial (ocular) clefts. Transverse Facial Cleft – Lateral Canthal(Ocular) - Right

Figure 14: No. X, Cranio Facio Cervical (Branchial) Clefts – 2. Skeletal – Cranial Cleft. A. Anterior A and B - Cleft of Fronto - Zygomatic & Temporal Bones - Right (Lateral Canthal region)
involving the soft tissue in between the eyelids in the medial canthal region and the nose [Table 1 and in Figures 1, 2 and 12]. The skeletal transverse cleft [Sk] runs in the transverse direction in the medial canthal region towards the midline, involving the nasal, ethmoid and lacrimal bones [Table 2 and in Figures 3 and 12]. The cleft may involve only the soft tissue structure or may involve both soft tissue and skeletal structures. The cleft may be unilateral or bilateral.

**Number X**

Number X is the lateral ocular Transverse facio cranial cleft.
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Figure 18: No. XIV (Branchial Cleft No. 4) Cranio Facio Cervical (Branchial) Clefts – 1. Soft tissue A. Anterior, group – 4. Cervical (Branchial) clefts. A, and B No. XIV (Branchial Cleft No. 4) – Bilateral

Figure 19: No. XIV, (Branchial Cleft No. 4) Cranio Facio Cervical (Branchial) Clefts -1. Soft tissue. A. Anterior, Group – 4. Cervical (Branchial) Clefts. A. No. XIV (Branchial Cleft No. 4) Bilateral, B. MRI is showing Bilateral clefts extending upto subcutaneous levels.

Figure 20: No. XV, Cranio Facio Cervical (Branchial) Clefts – 1. Soft tissue A. Anterior, Group – 5. Lower Median Facio-Cervical Clefts A. Median Cleft Lower Lip (& Cleft Lip – Left side), B. Median Cleft Lower Lip & Neck, C. Median Cervical Cleft, D and E Median Cleft Tongue, and F. Median Cleft Lower Gum
The soft tissue cleft \([S]\) runs in the transverse direction from the lateral canthal region towards the temporal region, involving the soft tissue in between the eyelid in the lateral canthal region and temporal region [Table 1 and in Figures 1, 2 and 13]. The skeletal transverse lateral ocular cranial cleft \([Sk]\) runs in the transverse direction in the lateral canthal region towards temporal region, involving frontozygomatic and temporal bones [Table 2 and in Figures 3 and 14]. The cleft may involve only the soft tissue structure or may involve both soft tissue and skeletal structures. The cleft may be unilateral or bilateral.

**Number XI**

Number XI is the First Branchial Cleft Anomalies. The external opening of this cleft is below the external auditory canal in the anterior border of the sternocleidomastoid muscle [Table 1 and in Figures 1, 2 and 15]. The internal opening is in the Tonsillar sinus or near the palatopharyngeal arch. In this Branchial cleft anomaly there may be only soft tissue involvement or a combination with cartilage clinically recognized as Branchial vestige. The cleft may be unilateral or bilateral.
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Figure 23: No. 1 B. Cranio Facio Cervical (Branchial) Clefts – 1. Soft tissue Group B. Posterior – Cervical, Lower Median – A and B. Posterior Cervical Cleft [Rachischisis] C and D. Posterior Cervical Encaphalocele

Figure 24: No. 1 A. Cranio Facio Cervical (Branchial) Clefts – 2. Skeletal-Bony B. Posterior Cranial

**Number XII**

Number XII is the Second Branchial Cleft Anomaly. The external opening of this cleft anomaly is at the junction of the upper two thirds and lower one third in the anterior border of the sternocleidomastriod muscle [Table 1 and in Figures 1, 2 and 16]. The internal opening is in the Tonsillar sinus or near the palatopharyngeal arch. This is a common variety of cleft anomaly. The cleft may be unilateral or bilateral.

**Number XIII**

Number XIII is the Third Branchial Cleft Anomaly. The external opening of this cleft anomaly is just below the external opening of the Number XII [Second branchial cleft
anomaly], in the anterior border of sternocleidomastriod muscle [Table 1 and in Figures 1, 2 and 17]. The internal opening is in the Tonsillar sinus or near the palatopharyngeal arch. The cleft may be unilateral or bilateral.

**Number XIV**

Number XIV is the Fourth Branchial Cleft Anomaly. The external opening of this cleft is in the anterior border of the sternocleidomastriod muscle near the sternoclavicular joint [Table 1 and in Figures 1, 2, 18 and 19]. The internal opening of this cleft is in the Tonsillar sinus or near the palatopharyngeal arch. This cleft anomaly may occur as a unilateral or bilateral cleft. According to Converse Plastic Surgery, Volume 5, the Fourth Branchial cleft anomalies have not been clinically demonstrated, despite a theoretical basis for their existence. But in our series one case of Bilateral Number XIV cleft [Fourth Branchial Cleft anomaly] [Figures 18 and 19] has been documented.

**Number XV**

Number XV is the Lower median facio cervical and tongue
cleft [Glossoschisis]. The cleft may be seen as a soft tissue cleft of the lower lip and tongue in the midline. The cleft may be seen in the skeletal structure i.e. mandible or cartilaginous structures like hyoid, thyroid, cricoid and trachea in the midline. The soft tissue cleft starts from the oral cavity in the midline of the lower lip, and runs in the downward direction in the neck, some time upto the sternal notch. The cleft may involve these soft tissues in varying degrees. The external opening of this cleft is in the midline of the neck and the internal opening is in the tongue [Table 1 and in Figures 1, 2 and 20]. The median cervical cleft as seen in Figure 20, represents remnants of the thyroglossal duct with external opening in the midline of the neck . The internal opening is in the foramen cecum of the tongue. The skeletal bony cleft occurs in the medium plane of the body of mandible [Table 2 and in Figure 21]. The skeletal cartilaginous cleft occurs in the median plane through Hyoid, Cricoid, Thyroid and Trachea [Table 2 and in Figures 21].

THE DESCRIPTION OF POSTERIOR CLEFTS IN THE HEAD AND NECK

The Posterior Cleft in the head and neck region occurs in the median plane anywhere between the vertex and the neck. Such posterior soft tissue and skeletal clefts in the head and neck regions are described as 'Craniorachischisis'. The cleft in the posterior region of the head is the "Cranioschisis" and is grouped under IA [Figure 22]. This cranial cleft is clinically noted as Posterior cranial-occipital encephalocele [notencephalocele] and meningomyelocele [Figure 22]. The cleft in the posterior region of the neck and further down is the "Rachischisis" and is grouped under IB [Figure 22]. This cervical cleft in the posterior region is clinically noted as a Posterior cervical encephalocele and meningomyelocele [Figure 23]. Usually the soft and skeletal tissue clefts in the posterior region of the head and neck region occur together. The cleft from the cranial region may extend to any level of the vertebral column. Such conditions are not usually compatible with life. The clefts in the Posterior aspect in skeletal structure occur in the occipital bone in the median plane [Figure 24] and in the cervical region through vertebrae in the median plane [Figure 25].

CLEFT LIP AND PALATE CLASSIFICATIONS

There are various Classifications described. Nagpur Classification advocated by Prof: C. Balakrishnan, is one of the classifications followed by many. The classification is as follows:

1) Group I - Cleft of lip [soft tissue]
   Group IA - Cleft of the lip and alveolus [soft tissue and skeletal combined].
2) Group II - Cleft of the palate only [soft tissue and skeletal]
3) Group III - Cleft of the lip and palate [soft tissue and skeletal combined]

All varieties of Clefts described in the Nagpur Classification can be mapped out assigning numbers in accordance with this proposed new classification.

Cleft lip gr I

Soft tissue cleft in the upper lip lateral to midline either as incomplete [Figure 26 A] or complete, unilateral [Figure 26 A] or bilateral [Figure 26 B], falls into Numbers 2 and 3 depending upon the anatomical location of the cleft.

Cleft lip gr IA

Cleft of the lip and alveolus, combined soft tissue and skeletal cleft, either unilateral or bilateral, skeletal cleft running through alveolus and the palatine bone shelf to a variable distance, medial or lateral to canine tooth [Figure 26 C].

Cleft palate gr II

Cleft of the soft and skeleton tissues - Palatal shelf, usually in the midline falls into Number I [S and Sk] [Figure 26 D].

Cleft gr III

Cleft of the Lip and Palate. Combined soft and skeletal tissue cleft groups described above may be unilateral [Figure 26 E] or bilateral [Figure 26 F] (falls into numbers 2 and 3). Cleft lip and palate of other Classifications can also be mapped out and numbered in accordance with the new classification taking into account the anatomical locations.

CONCLUSION

This paper aims at presenting a simple classification of all the clefts in the head and neck region. Duplication in numbering defects is avoided. The clefts run from the central point of reference the oral cavity, towards the facial,
cranial and neck region, and numbered accordingly. The clefts occur either as isolated or in combination of soft tissues or skeletal structures in varying degrees. Severe degrees of clefts are probably not compatible with life. The combination of several clefts and associated malformations are a possibility. The new classification offers to eliminate confusing terminology.

1) Cleft lip and palate cases are to be further examined and investigated to rule out the presence of other hidden clefts.

2) A system is developed for clinical evaluation, surgical exploration, mapping of anatomical landmarks in soft tissue and skeletal tissues as the internal relations among the cranium, face and neck are well established.

Wide differences of approach of Plastic and Reconstructive Surgeons in the management of cleft lip and palate with that of other clefts needs to be narrowed down, as the new classification broad bases the assessment and management and makes it more rational.

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