IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 17, Issue 8 Ver. 1 (August. 2018), PP 01-05
www.iosrjournals.org

Congenital Anomalies in Otorhinolaryngology

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Abstract: *A number of factors like genetic, intrauterine infection and maternal nutritional deficiencies are involved in complex development of brachial apparatus and cochlea formation. A clinician should examine the child thoroughly for other organ developmental anomalies.. The counseling and children management shouldbe considered hand by hand.*

Keywords: *brachial apparatus, cochlea, developmental anomalie*s

Date of Submission: 23-07-2018 Date Of Acceptance: 06-08-2018

I. Introduction

All children are special but children with congenital anomalies are more special because of their need for special medical and surgical interventions.

The otorhinolaryngological developmental anomalies are predisposed by many genetic factors, intrauterine viral infections, and maternal nutritional deficiencies. The congenital otorhinological anomalies can be associated with other systemic maldevelopments. The E.N.T. surgeon must look for associated anomalies of cardiovascular, gastrointestinal tract and renal malformations. Parents of suspected children should be counseled properly to investigate other systemic anomalies and managed accordingly.

II. Aim And Objectives

To overview the occurrence and presentation of congenital otorhinological anomalies.To study association between E.N.T. anomalies and other systemic involvementto study available surgical modalities and devices in management of congenital anomalies.

III. Materials and Methods

Total 136 cases with congenital anomalies in E.N.T., who presented in different departments of Government medical college and Sir. T. hospitalBhavnagar during period between January 2017 to May 2018 were included in this prospective study.

The patient examined thoroughly and evaluated for otorhinolaryngological and other systemic abnormalities. For earrelated developmentalanomalies history includes decreased hearing, deformity of external pinna or External auditory canal, pre auricular tags, pre auricular sinus and accessory tragus. The hearing assessment done with startle reflex, Tuning fork tests, free field audiometry, Oto-acoustic emissions,BERA test, ASSR test. MRI in some patient to rule out cochlear maldevelopments.Nasal developmental anomalies are examined and managed surgically. Neonates with respiratory distress are investigated by passing 6 or 8 French gauzeRyle's tube to rule out choanal atresia or stenosis. The cases with bilateral congenital choanal atresia are managed by various modalities of surgery. The cleft including lip and palate are examined, classified and managed initially with various dental apparatus and thereafter by surgery. The cleft patient should be evaluated for ear problems and congenital heart diseases. The patient with ankyloglossia are identified classified and tongue tie released surgically and Post operatively managed with speech therapy. The patients with noisy breathing must be investigated for laryngeal examination to rule out structural anomaly which may shows omega shaped epiglottis. All neck triangles are examined for congenital anomalies like branchial cyst and fistula, thyroglossal cyst and fistula, lymphangioma, congenital torticollis and sternocleidomastoid tumor.All patients are investigated with routine investigations like complete blood examination, renal function test, serum electrolytes, HIV, HBsAg.Special investigation like thyroid function test in thyroglossal cyst and congenital hypothyroidism, thyroid scan for thyroglossal cyst, chest x ray PA view, 2D echo, USG KUB, USG abdomen, HRCT/MRI temporal bone, CT PNS, CT neck, USG neck, sinogram, Fistulogram, FNAC done.

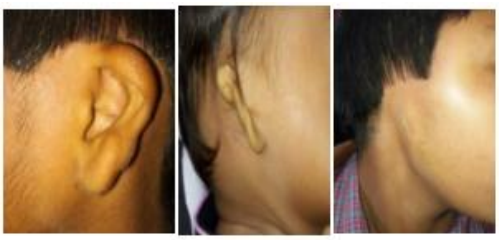
IV. Observation and Discussions

Out of the total 136 cases, 74 (54.41 %) were male and 62 (44.11 %) female.In year 2002, Sinha V, Johri S, and George A have done study of 100 patients of otorhinological developmental anomalies. According

to that study 64% male and 36% female were involved.[1] Maximum of patients presented in first decade of life which is corresponding with finding of this study records.[1]

NO.	ANOMALY	NUMBER OF CASES (%)	MALE CASES	FEMALE CASES
1.	Pinna and E.A.C. anomalies	11 (8.08%)	06 (4.41%)	05 (3.68%)
2.	Pre auricular sinus	03 (2.2%)	02 (1.74%)	01 (0.74%)
3.	Accessory tragus	02 (1.74%)	0 0	02 (1.74%)
4.	Pre auricular tag	05 (3.74%)	02 (1.74%)	03 (2.21%)
5.	Cleft anomalies	16 (11.67%)	09 (6.61%)	07 (5.14%)
6.	Congenital SNHL	63 (46.32%)	37 (27.20%)	26 (19.12%)
7.	Choanal atresia	04 (2.94%)	01 (0.74%)	03 (2.2%)
8.	Thyroglossal cyst	02 (1.74%)	01 (0.74%)	01 (0.74%)
9.	Thyroglossal fistula	02 (1.74%)	0 0	02 (1.74%)
10.	Brachial fistula	01 (0.73%)	01 (0.74%)	0 0
11.	Laryngomalacia	13 (9.55%)	07 (5.14%)	06 (4.41%)
12.	Tongue tie	07 (5.14%)	04 (2.94%)	03 (2.21%)
13.	Wry neck	04 (2.94%)	03 (2.21%)	01 (0.74%)
14.	Congenital facial weakness	02 (1.74%)	0 0	02 (1.74%)
15.	Depressed nasal septum	01 (0.73%)	01 (0.74%)	0 0
	TOTAL	136	74 (54.41%)	62 (45.59%)

Total 11(7.35%) patients have pinna abnormalities, out of which 5 patient have microtia, 1 patient have bilateral anotia, 2 patients of bat ear, 1 case of darwin tubercle, 2 cases of pinna sinus. Out of these patients,two patients are associated with total external auditory canal atresia.



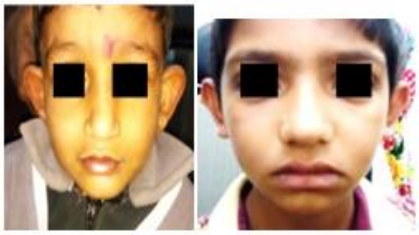
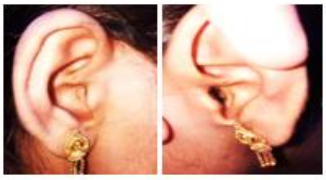
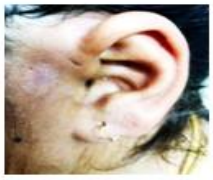


Fig.1(a),(b) : Microtia Fig.2 Anotia Fig. 3 (a) : Bilateral bat ear; (b): Bilateral darwin's tubercle

In our study three (2.2%) patient of bilateral preauricular sinus were recorded. In our study mean age of presentation for pre auricular sinus is 27 year. [2]





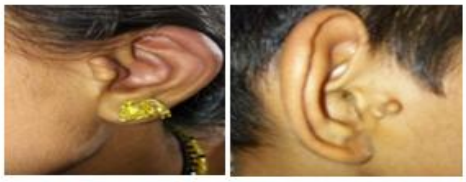


Fig. 4 (a),(b) : Bilateral pinna sinus (c) left ear pre auricular sinus; (d) pre auricular tag ; (e) accessory tragus

Two (1.47%) female patients have left sided accessory tragus and five (3.67%) cases of right sided pre auricular tag were identified with ratio of 3:2 for male to female.

V. Cleft Deformities

TYPE OF ANOMALIES	SIDE OF ORAL CAVITY AFFECTED	NO. OF CASES
SUBMUCOSAL CLEFT PALATE	MIDLINE	4
CLEFT LIP	LEFT	2
INCOMPLETE CLEFT PALATE	MIDLINE	2
	LEFT	2
	RIGHT	0
COMPLETE CLEFT PALATE	LEFT	2
CLEFT PALATE + LIP	LEFT	1
CLEFT PALATE + LIP + NOSE	BILATERAL	1
	RIGHT	1
	LEFT	1
TOTAL		16

Table 2: various cleft anomalies enumerated in table including number of cases.[1][3]

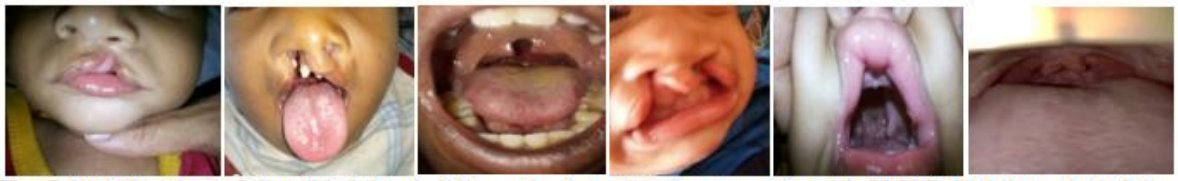


Fig. 7 (a) left side cleft lip; (b) right cleft lip and palate; (c) absent uvula with SMCP (d) bilateral cleft lip; (e) midline complete cleft palate; (f) bifid uvula with SMCP

Total 63(46.32%) patientswere noted with congenital sensory hearing loss. In our study, average age of diagnosis is 3.73 years.All affected bilaterally. The prelingual patients were encouraged for cochlear implant surgery and BAHA hearing aid. Two patients of waardenburg syndrome with congenital sensory neural hearing loss, isochromia and heterochromia of iris, dystrophia canthorum were noted.[1][4]

Fig. 10 (a) isochromia of iris; (b) heterochromia of iris with dystrophica canthorum and congenital SNH

Choanal atresiacan be perfectly described by "Birth without breath." Total four (2.94%) patients of choanal atresia were noticed. In our study male to female ratio is 1:3.Two patients of each bilateral and unilateral choanal atresia were noted and managed. [1][5][6][7]

An anomalies related to persistent thyroglossal duct were detected during period of 5 year to 9 year of age and in form of subhyoid variety. There were two patients each of thyroglossal cyst and fistula were notified during study period.All patient have normal thyroid function test, thyroid scan study. All four patient were managed successfully in Government medical college and Sir T. hospital Bhavnagar.[8]

Fig. 11 (a), (b): thyroglossal fisula

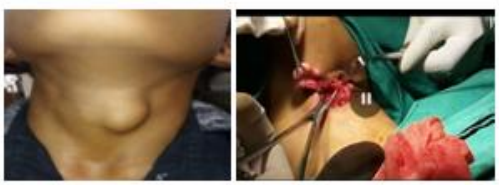


Fig. 12 (a),(b): Thyroglossal cyst

Onemale patient of right side neck branchial fistula was found and investigated with fistulogram, ultrasonography. Patient were operated in sir. T. hospital Bhavnagar with stepladder technique. The tract was extended up to right side tonsillar fossa region was successfully excised under general anaesthesia.[9]

Fig. 13: branchial fistula

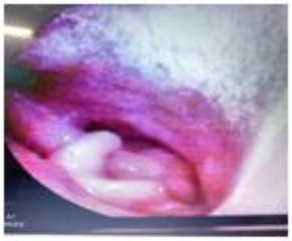


Fig. 14 laryngomalacia

The classical omega shaped epiglottis was examined by rigid 70* endoscope in 13 (9.55%) cases of noisy breathing. The noisy breathing sounds were reduced with prone position. In our study, age of presentation was between 1st to 4th months of age. All parents counseled and reassured.[10]

Seven (5.14%) patients of ankyloglossia were recorded.In our study, there were four male and three female. Three case of grade 1 tongue tie, another three cases were grade 2 and one with grade 3 were found during study. All patients were investigated with routine blood investigations and underwent for surgical excision and suturing with absorbable polythread suture material.[11]

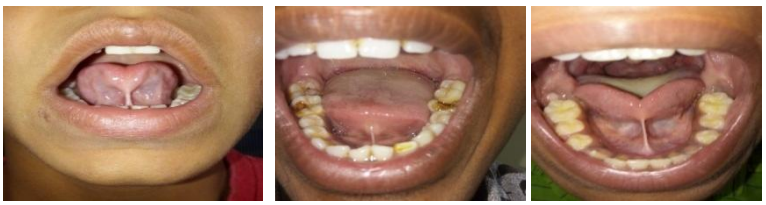


Fig 15. Ankyloglossia (a) grade;1 (b) grade 2; (c) grade 3

Total four (2.94%) patients were presented with congenital torticollis in period of 4th to 8th week of age with exception of one 6 year female with right side torticollis was presented as neglected case of wry neck. Two male patients of left sided sternocleidomastoidtumor and two female patients of right sided shortening of sternocleidomastoid muscle were notified.[12]

Fig. 16: (a) congenital torticollisFig. (b) Right congenital facial weakness (c) depressed nasal septum

Two (1.47%) neglected female cases of right congenital facial weakness were notified at the age of 16 year and 33 years.[13]

One male patient of 4 months old male child have depressed nasal septum with dystrophia canthorum, right renal agenesis, and patent ductus arteriosus.

VI. Conclusion

External ear deformities wereseen in 20 (14.7 %) cases.

Congenital anomalies related to inner ear presented with congenital S.N.H.L. seen in 63 (46.32 %). Which account for near half of the patients.

Cleft deformities seen in 16 (11.67 %) patients. Out of which 9 (56.25%)were male and 7 (43.75%) were female. Four (25%) cases associated with congenital heart disease and 6 (37.5%) patients were associated with ear problems. In our study, 4 (2.94%) cases identified as choanal atresia. Male: female ratio is 1: 3. In our study, bilateral choanal atresia found in 50% cases managed with transpalatal and transnasal approaches.While postoperatively, Vikas Sinha stenting method was used for patency.

Anomalies related to thyroglossal duct found in four (2.94%) cases. Two patients of subhyoid thyroglossal cyst were managed surgically. One (0.73 %) case of branchial fistula found which was managed surgically by stepladder method.13 (9.55%) cases of laryngomalacia were identified. Ourstudy shows slight predominance of male (7 cases) rest of 6 cases were female.

We found seven cases of tongue tie. Out of which four were male and three were female. We found three cases of grade one ankyloglossia and grade 2 ankyloglossia and rest of one patient of grade 3 ankyloglossia.

We found four (2.94%) case of congenital torticollis, 2 (1.47%) cases of congenital facial weakness, one (0.735%) case of congenital dacryocystitis and depressed nasal septum.

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