CO-EXISTENCE OF OBLIQUE PINNAE AND CONGENITAL HEART DISEASE

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ABSTRACT

The paper reports a syndrome in which oblique placement of one or both the pinnae on face was found to co-exist with congenital -anomalies of heart like VSD, PDA and Tetralogy of allot, etc. Although clinically discernible in 90% of the subjects, objective evidence of obliquity of pinna was documented by photogrammetry. The values for the ear inclination in subjects categorized clinically as normally placed pinna were 7.9° ± 339° with range 2°-17". In contrast, subject where the pinna was clinically categorized as oblique had mean AEI 16.5°±5.81 ° with a range 5°-33°.

Eighteen of the 20 subjects with oblique pinna were demonstrated to have some congenital anomaly of heart as indicated by clinical and echocardiographic examination. In contrast, in subjects with normally placed pinna only 3 out of 34 possessed clinical heart defects. Presence of oblique pinna indicates a thorough search for undetected heart defects.

Keywords: *Oblique pinna, Congenital heart disease.*

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Abnormalities of the shape and size of external ears have been reported to be associated with gross abnormalities of internal organs. These include CHARGE syndrome(l,2). Langer-Giedion syndrome, chromosomal anomalies like XXY, monosomy G, Down syndrome, Crouzon, Apert, Cranio-oculodental, Treacher Collin's, Bixler and Goldenhar, Klippelfeil and Fanconi, *etc.*(3-5). The abnormalities encountered in above named syndromes include misshapen pinna, poor development of ear cartilage and its lower placement relative to intercanthal line.

A recent computer search of literature failed to reveal any described relationship between obliquity of the pinna and congenital heart defects. We describe below 20 cases of a syndrome which we believe has not been described earlier in which obliquity of pinna (posterior displacement of the vertical axis) was often associated with congenital heart defects like Ventricular Septal Defect (VSD) and Patent Ductus Arteriosus (PDA). Tetralogy of Fallot and Pulmonary Stenosis were other congenital malformations detected in a few cases.

Material and Methods

The material comprised of 20 children (age range: 3 months to 11 years) of both sexes whose pinnae appeared to be obliquely placed on clinical examination. Thirty four children (age range: 1-16 years) with pinnae that appeared normal on clinical examination were studied to obtain values for "angle of ear inclination".

All subjects had visited the Out-Patient Clinics of the Nehru Hospital of the Postgraduate Institute of Medical Education and Research, Chandigarh, in the last three years. A complete clinical examination of cardiovascular system was carried out in

children of both groups, echocardiography was performed in children who were suspected to have congenital heart disease on the basis of one or more clinical signs to demonstrate the anatomy of cardiac defects.

Profile photograph of the left side of the face of all study subjects was taken including the entire helical rim of left ear, while keeping the head in the Frankfurt Horizontal (FH) plane. Points, orbitable (o) and tragion (t) were marked with a colored skin marking pencil on subject's face so as to determine the exact position of FH plane before taking the photograph of the subject. Help of an attendant was sometimes necessary for fixing the position of head of an infant in FH plane. All measurements were taken on the left pinna in conformity with the accepted practice, though on clinical examination both pinnae were found to be oblique.

The angle of ear inclination (AEI) was directly measured on the photographs fixed on a large white paper sheet. The angle subtended between longitudinal axis (AB) of ear and vertical plane (BC) of the head or face with head positioned in FH plane was considered as angle of ear inclination. The longitudinal axis of ear (AB) was determined as per technique given by Farkas(6,7), while vertical axis (BC) was represented by a perpendicular drawn from point B on the Frankfurt Horizontal plane (0 to t line) *(Fig. 1)*. The angle was measured with the help of a protractor upto the accuracy of 1° (one degree). All the measurements were recorded by a trained anthropometrist (AKB). The difference in two measurements taken by the same observer (AKB) was less than one degree.

Results

The AEI in children who had normal looking ears as compared to those in whom the pinna were considered to be oblique on inspection is shown in *Table I.* There were 34 children who had been categorized as possessing normal ears on inspection. Their cardiovascular- system did not show any abnormality on clinical examination. Echocardiography was not carried out. Three of them were subsequently shown to have AEI exceeding 12° on photogrammetry showing thereby that obliquity of ears can be missed occasionally. In the group of 20 subjects with obliquely placed pinna on clinical examination and AEI values exceeding 12°, clinical and echocardiographic evidence of congenital heart disease was present in 18 subjects (90%).

The mean angle of ear inclination was 16.5° ± 5.81° with a range of (5°-33°) for children with oblique looking pinna, whilst AEI values recorded in children with normal looking pinna was of the order of 7.9° ± 3.39° (range 2°-17°).

Discussion

The oblique placement of the pinna was clinically discernible. Infact, all our subjects were picked up in this way. Measurement of angle or ear inclination was resorted to only for the purpose of providing objective evidence for the report. In children with normal looking ear, AEI exceeding 12° was observed in 3 of the 34 subjects (8.8%) on photographic measurement, thus clinical observation is adequate to identify oblique placement of pinna in more than 90% of the subjects.

The mean AEI in heart defects was more than two times as compared to children who had normal ears and normal hearts. The values for AEI reported by Farkes(7) ranged between 6.3°-35.6° (mean 20°) in normal girls and 9.3°-31.2° (mean 21°) in

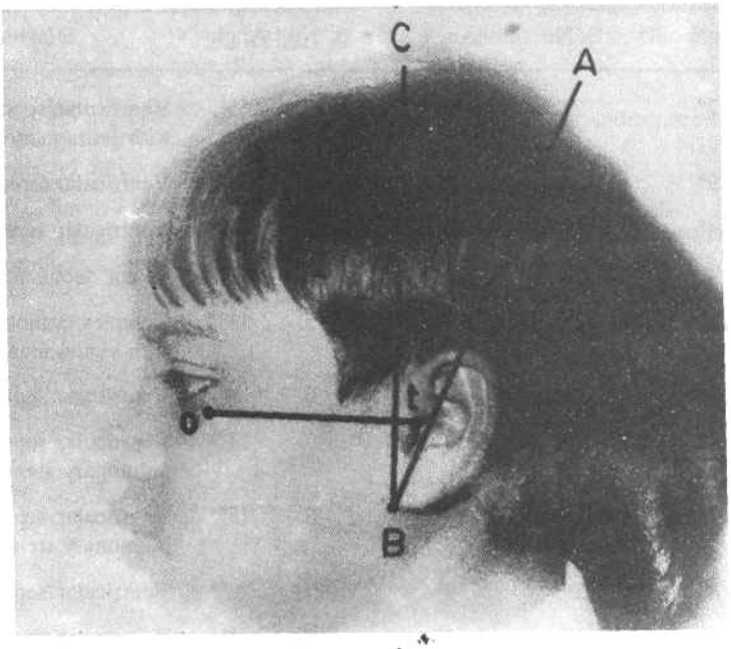

Fig. 1. The measurement of angle of ear inclination

boys. However, he had not examined the heart and has therefore, not commented on any co-existing cardiac abnormality. Our data clearly indicate the presence of congenital heart disease on the basis of clinical and echocardiographic examination in 18 of the 20 subjects having AEI more than 12°.

During embryologic development of pinna, posterior displacement in the vertical plane of the face is well described. The external ear appears in the second month in the region around the first pharyngeal groove. At about the same time (between 32nd and 46th day), interventricular septum is also developing(8). It is possible that the same agent/stimulus acting at this vulnerable period distorts the development of both the regions simultaneously. Our data suggest that any child whose pinna appears obliquely placed on clinical examination should be carefully investigated for co-existence of unrecognized congenital heart anomalies. Considering that we have encountered 20 subjects with this syndrome in the last three years, the correlationship may not be uncommon even though it has remained undescribed so far.

TABLE I–*Angle of Ear Inclination (AEI)*

Children with normal pinna (n = 34)				Children with oblique pinna (n = 20)		
S. No.	Angle	S. No.	Angle	S. No.	Angle	Diagnosis
1.	2°	21.	6°	1.	5°	Ventricular septal defect with pulmonary stenosis
2.	9°	22.	7°	2.	33°*	Ventricular septal defect (VSD)
3.	4°	23.	10°	3.	16°*	Ventricular septal defect
4.	4°	24.	10°	4.	13°*	Patent ductus arteriosus (PDA)
5.	12°	25.	7°	5.	13°*	Complex cyanotic heart disease with ventricular septal defect
6.	17°*	26.	8°	6.	8°	Ventricular septal defect
7.	9°	27.	10°	7.	13°*	Ventricular septal defect with pulmonary stenosis
8.	7°	28.	10°	8.	15°*	Ventricular septal defect with pulmonary stenosis
9.	8°	29.	9°	9.	18°*	Ventricular septal defect
10.	3°	30.	6°	10.	13°*	Ventricular septal defect
11.	11°	31.	10°	11.	16°*	Tetralogy of Fallot
12.	15°*	32.	8°	12.	25°*	Ventricular septal defect
13.	3°	33.	13°	13.	14°*	Transposition of great arteries with pulmonary stenosis
14.	2°	34.	7°	14.	15°*	Patent ductus arteriosus
15.	5°			15.	17°*	Ventricular septal defect
16.	8°			16.	19°*	Ventricular septal defect
17.	6°			17.	19°*	Ventricular septal defect
18.	6°			18.	19°*	Ventricular septal defect
19.	7°			19.	19°*	Ventricular septal defect
20.	10°			20.	17°*	Ventricular septal defect
			Mean ± SD			Mean ± SD
			7.9° ± 3.39°			16.5°±5.81°

* AEI measures >12°.

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