The Premaxillary Triangle

Clue to the Diagnosis of Cleft Lip and Palate

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Objective. The prenatal detection rate of cleft lip and palate is low, especially in low-risk patients who undergo targeted sonography. The reason is that evaluating surface anatomy is relatively difficult and requires operator expertise. Our purpose was to describe a technique to improve the diagnostic accuracy of facial clefts (lip and palate) and to assess the feasibility of including this technique as part of standard protocol during targeted imaging. ***Methods.*** A prospective study was done during 2000 through 2002 to evaluate the accuracy of the "premaxillary triangle (PMT) sign": a new sign to diagnose unilateral cleft lip and palate in women referred for prenatal sonography at our center. Patients with only isolated unilateral cleft lip and palate and cleft lip were included in this study. Before this, all examiners were trained to image the PMT. The images were reviewed by a senior consultant. It was later decided to include this sign as part of the protocol of targeted sonography done between 18 and 22 weeks in our institution. However, depending on the fetal position, the PMT was documented even in patients referred for the first time in late second and third trimesters. ***Results.*** Twenty-nine cases of isolated facial clefts were diagnosed during the study period, of which 2 had unilateral cleft lip and 27 had unilateral cleft lip and palate. The PMT sign was absent in all cases of unilateral cleft lip and palate but was present in 2 cases of isolated cleft lip without cleft palate. ***Conclusions.*** The PMT sign can be easily incorporated into targeted sonography at 18 to 22 weeks' gestation. Its inclusion would help in increasing the detection rate of unilateral cleft lip and palate. It may also be potentially used for differentiating between isolated cleft lip and cleft lip and palate, which helps in better prenatal counseling.
Key words: cleft lip; cleft lip and palate; premaxillary triangle; triangle sign; unilateral lesion.

Abbreviations
PMT, premaxillary triangle

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The prenatal detection rate of cleft lip and palate is low, especially in low-risk patients who undergo routine targeted scans, although the incidence of facial clefts is approximately 1 per 500 to 1000 live births.[1] Stoll et al[2] and Clementi et al[3] reported an increase in the detection rate from 5.3% in 1979 through 1988 to 26.5% in 1989 through 1998. The reason for the poor accuracy is that the anatomic structures inside the fetus are identified more easily than surface anatomy. In a retrospective study, Cash et al[4] reported 67% sensitivity, and in a prospective study, Wayne et al[5] reported 75% sensitivity in diagnosing isolated facial clefts in a routine obstetric population scanned between 18 and 23 weeks. Three-dimensional imaging has not only increased the detection rate but also helped in delineating precisely the bony extent of facial clefts.[6] Until recently, imaging of the face was not included as part of targeted sonography per-

formed between 18 and 22 weeks.[7,8] However, operator expertise is essential to diagnose facial abnormalities, especially between 18 and 20 weeks. In this report, we describe a new sonographic method of evaluating the fetal face to identify facial clefts between 18 and 22 weeks. This technique involves identification of the "premaxillary triangle" (PMT) which, to our knowledge, has hitherto not been described.

Materials and Methods

The 3 standard views in which the face is imaged are coronal, axial, and sagittal views. Facial clefting is visualized clearly in the axial view of the face. However, the coronal view of the face is relatively easier to obtain when compared with the axial and sagittal views. In practice, we have observed that the coronal view of the face can be obtained from the biparietal diameter plane of the head by moving the transducer toward the anterior aspect of the fetus with a 90° rotation. Also, the lesser amount of soft tissue in the second trimester makes the bony landmark more pronounced in the coronal view of the face.

From the coronal view of the face, the transducer is tilted to obtain the 2 angulated nasal bones, forming an inverted V shape. The V is completed into a triangle by imaging the premaxillary bone, which forms the base of the V. We have named this triangle the PMT. The symmetrically placed orbits should not be visualized because the nasal bones as an inverted V are not visualized in this posterior coronal section.

In our study, it was observed that the PMT could be imaged in fetuses as early as 14 weeks' gestation, without an increase in the time taken for completing the scan. Hence, it was decided to include this in the protocol of the second-trimester targeted scan. An attempt was made to image the PMT in the late second and third trimesters also. A coronal section of the face showing the normal PMT at varying weeks' gestation is shown in Figure 1, A–D. In the late second and third trimesters, the development of the incisor tooth buds is seen as parallel hyperechoic lines at the level of the premaxilla (Figure 2). Absence of the PMT is diagnostic of cleft lip and palate (Figure 3, A and B).

During a 6-month period from June 1999 to December 1999 in our institution, all the examiners were trained to image the PMT during antenatal evaluation. The systems used were HDI 5000 and Envisor (Philips Medical Systems, Bothell, WA), Voluson Xpert (GE Healthcare, Milwaukee, WI), and SSD-3500 (Aloka Co, Ltd, Tokyo, Japan). A standard convex probe with a 3.5-MHz frequency was used in all cases. In addition to documentation of the PMT, the axial and coronal sections of the lips and the axial section of the maxilla and mandible were also documented. The images of the PMT were then reviewed by a consultant (S.I.).

A prospective study was done during January 2000 through December 2002. Follow-up of all the patients who underwent antenatal sonography in whom the PMT was documented was difficult because ours is a referral sonography center and patients give birth in different parts of our country. Hence, to strengthen our study, we retrospectively collected the follow-up of 500 women, referred to us for targeted scans between 18 and 22 weeks for screening sonography from a single hospital, in whom the PMT had been documented. We chose this gestational age group because the purpose of this study was to assess the feasibility of including the PMT in the protocol of targeted imaging.

Results

During the January 2000 through December 2002, a total of 34,211 obstetric scans were performed. A total of 68 facial clefts were identified (0.2%). Thirty-nine (57%) of the 68 cases were seen with other system anomalies and hence were excluded from the study. Isolated facial clefts were seen in 29 (43%) of 68.

Of the 29 cases of isolated facial clefts, 2 were isolated cleft lip, and the remaining 27 cases had cleft lip and palate. The gestational age at referral varied between 16 and 36 weeks, of which 9 cases were between 16 and 24 weeks and rest were between 24 and 36 weeks. The diagnosis of a facial cleft was missed in 3 cases referred before 24 weeks. In case 1, there was an isolated cleft lip, which showed a normal PMT (Figure 4, A–C). In case 2, there was improper documentation of the triangle sign (Figure 5), and in case 3 (Figure 6), the absence of a PMT was overlooked. However, all the 3 patients had another scan at 30, 26, and 28 weeks, respectively, and the diagnoses of cleft lip in the first case and cleft lip and palate in the other 2 cases were made. The reasons for the additional scans were as follows: in case 1, the patient had vaginal bleeding; in case 2, because it was an in

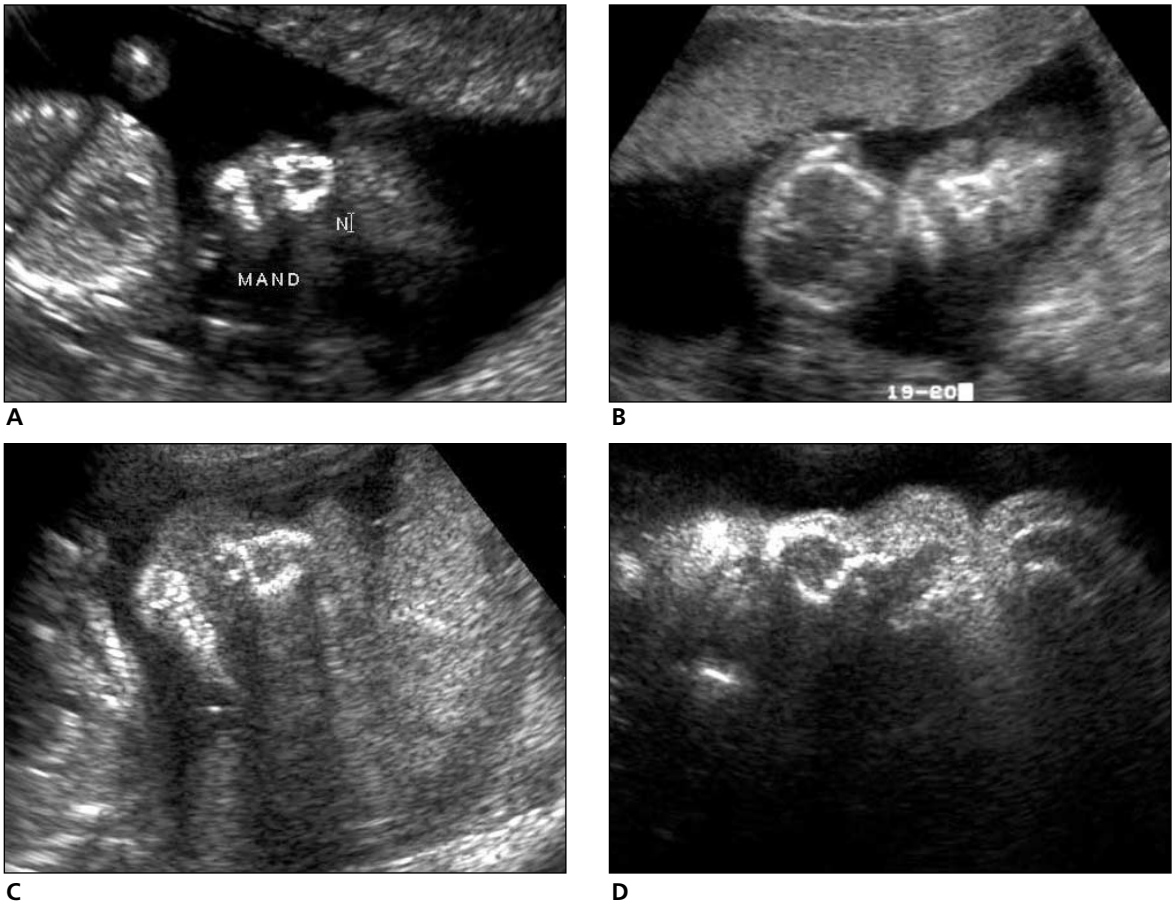


Figure 1. A, Coronal section of the face showing the normal PMT (N) at 12 to 13 weeks. MAND indicates mandible. **B**, At 19 weeks. **C**, At 25 weeks. **D**, At 36 weeks.

vitro fertilization conception and maternal obesity was present, we had advised a follow-up scan at 24 weeks, but the patient reported at 26 weeks; and in case 3, the patient insisted on having an additional scan at 28 weeks because she had a previous child with a unilateral cleft lip and palate at which time the anomaly was diagnosed. This patient opted for termination of pregnancy.

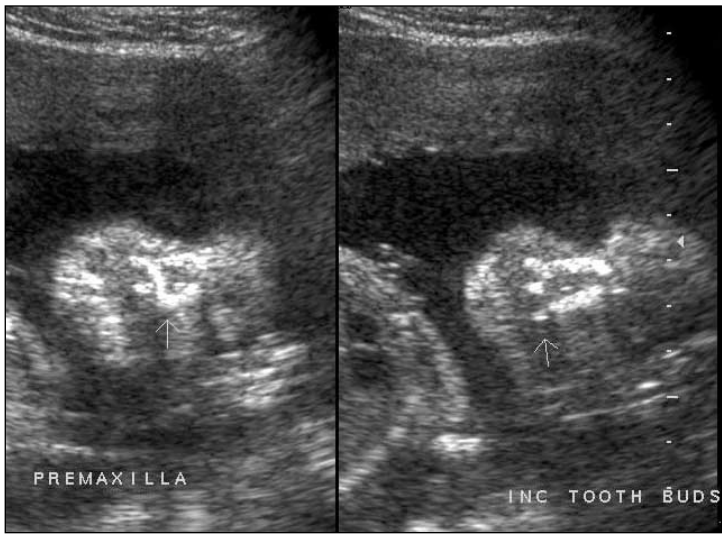
Twenty-eight of the 29 patients were followed postnatally, and all the neonates had postnatal surgical correction.

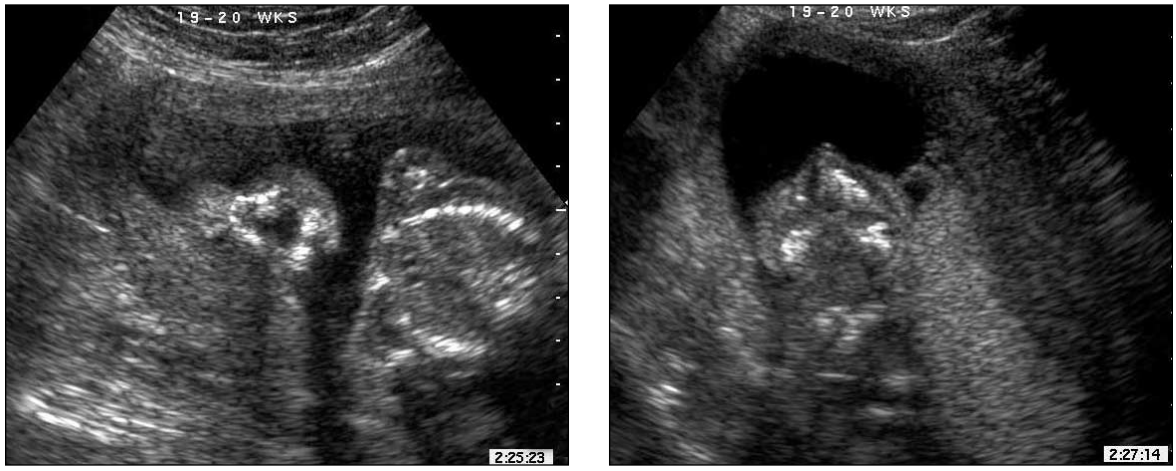
In the 500 cases that were referred from the single hospital, there was no case of isolated unilateral cleft lip or cleft palate, and in all the cases, the PMT had been documented.

Discussion

Nyberg et al[9] proposed a sonographic classification of cleft lip and palate. Christ and Meininger[10] were the first to diagnose cleft lip and palate ante-

Figure 2. Normal premaxilla and incisor (INC) tooth buds (arrows).



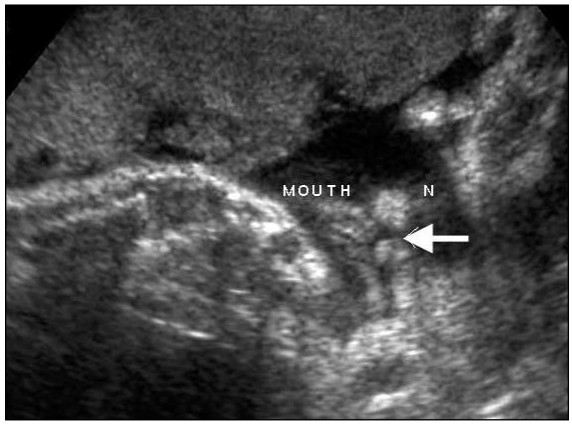


A B

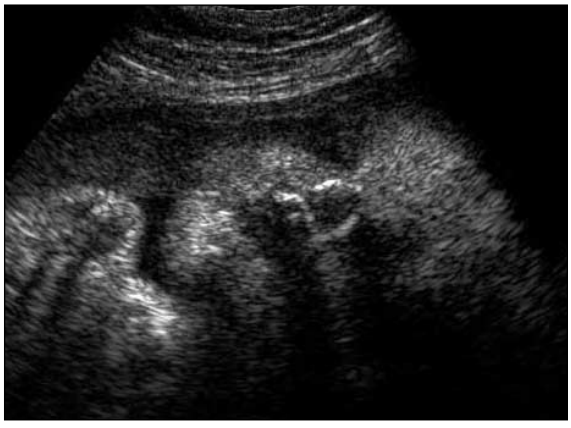
Figure 3. A, Coronal scan showing the absence of the PMT. **B**, Axial scan in the same fetus, which confirms the diagnosis of a unilateral cleft lip and palate.

natally in the third trimester. Facial clefts are suspected antenatally when a soft tissue protrusion is seen in the profile view of the face. Babcock et al[11] and Babcock and McGahan[12] described a sonographic method to identify midfacial abnormalities. They concluded that if a facial cleft is detected in the coronal plane, an axial section of the fetal upper lip and tooth-bearing alveolar ridge helps accurately differentiate between an isolated cleft lip and a cleft lip and palate and also detect unilateral versus bilateral defects. The identification of premaxillary soft tissue protrusion in the midsagittal section is a pointer for a bilateral cleft lip and palate.[13,14] However, a sagittal section of the face may not reveal the presence of a unilateral cleft lip if the defect is on the contralateral side. Proper recognition and imag-

A



B



C

Figure 4. A, Normal PMT at 20 weeks. **B**, Isolated cleft lip (arrow) with intact palate at 30 weeks. N indicates nose. **C**, Normal PMT at 30 weeks.

ing of the premaxilla helps in the diagnosis of an associated cleft palate in patients with the diagnosis of a unilateral cleft lip. The premaxilla is easier to obtain in the coronal view along with the nasal bone landmarks, especially in early weeks of gestation. Identification of the PMT does not exclude an isolated cleft lip, and this may be missed in the early weeks of gestation. An isolated cleft lip is diagnosed with greater accuracy in the third trimester because of better soft tissue delineation.[15]

The hard palate has the premaxillary portion and the palatine process. The palatine process is derived from the maxillary process. The premaxilla, otherwise called the primary palate, is formed from the frontonasal process. Formation of the palate is by fusion of both the palatine process and the premaxilla. The junction of the primary and secondary palate is called the incisive foramen. The cleft can involve the premaxilla or extend along the fusion line of the secondary palate. The involvement of the premaxilla leads to the absence of the triangle.

Figure 5. Posterior coronal image showing the orbits. The PMT is not visualized in this view.

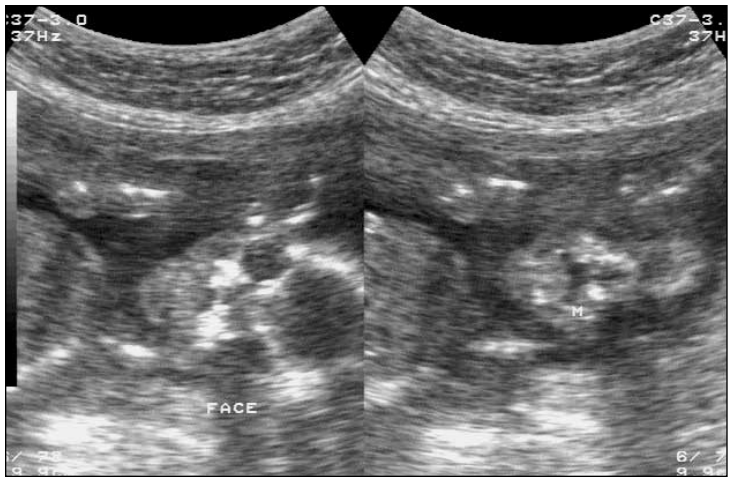


Figure 6. Right, Coronal view showing the absence of a base of the PMT. Left, Posterior coronal view with both orbits. M indicates mouth.

In conclusion, the inclusion of the PMT in the protocol of a targeted scan helps in increasing the detection rate of cleft lip and palate. Absence of the PMT is a strong pointer of a unilateral cleft lip and palate, which is confirmed by evaluating the soft tissues and the maxilla in the axial plane. The PMT may also be potentially used for differentiating between an isolated cleft lip and a cleft lip and palate. This precise information will help in counseling parents about the implications of the abnormality. Imaging the PMT, although simple and not time-consuming, requires a period of training for proper imaging and documentation to improve accuracy. Further studies are required to accurately document the sensitivity and specificity of this finding.

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