

MEDIAN CLEFT OF THE UPPER LIP IN ASSOCIATION WITH BILATERAL HEXADACTYLY AND ACCESSORY TOES

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Median clefts of the upper lip are rare. A case is described in which a median cleft of the upper lip was associated with symmetrical bimanual hexadactyly and bilateral accessory toes. A thorough search of the literature has revealed two similar cases and it is suggested that the association is sufficiently constant to form a recognisable syndrome.

CASE REPORT

A 26-year-old male nurse of Indian descent presented with a median notch of the upper lip and six digits on each hand and foot. There was no history of maternal illness during pregnancy and no other members of his family were similarly affected.

The notch (Fig. 1) was placed exactly in the midline of the upper lip vermilion and extended only as far as the red margin. The orbicularis muscle fibres were interrupted in the midline with absence of the characteristic philtrum complex. This, however, was adequately camouflaged by a moustache. The lip, columella and nostrils were otherwise normal. The frenulum, alveolus and teeth were also normal.



FIG. 1. View of upper lip to show midline cleft.

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The extra fingers were symmetrically placed on the ulnar borders of the hands and there was a complete ray for each extra digit (Fig. 2A). They were of normal morphology but were smaller than the adjacent normal digits. The extra toes were also on the lateral sides of the feet (Fig. 2B). On the left side the two smallest toes had separate phalanges but shared the same metatarsal bone. On the right side the fourth and fifth toes had separate but abnormal phalanges and were syndactylised. The sixth toe was normal in structure and appearance.

X-rays of the chest demonstrated normal ribs. Chromosome studies revealed a normal genotype.

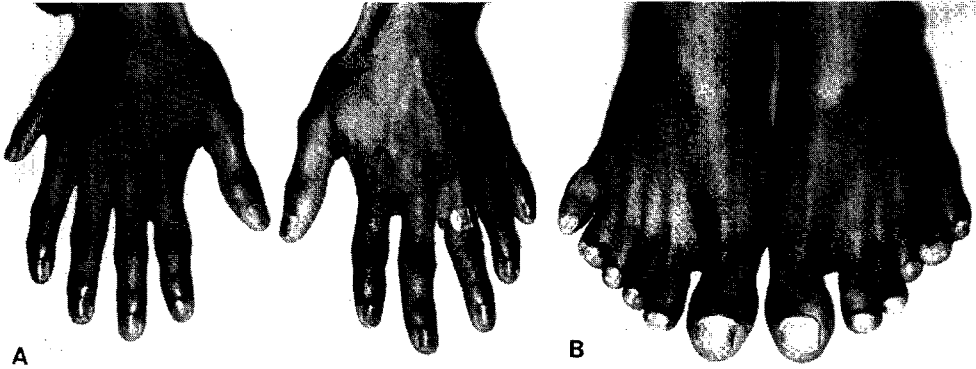


FIG. 2A. Both hands show polydactyly and incomplete webbing in the interdigital clefts. B. Both feet show accessory toes and web malformations.

TREATMENT

The notch of the upper lip was corrected by V-Y advancement as for the "whistle tip" deformity, combined with restoration of the continuity of the orbicularis muscle fibres. At a later date a Z-plasty to the vermilion was carried out (Fig. 3). The extra digit on the left hand and the lateral digit on



FIG. 3. Postoperative view of the completed repair of the cleft of the upper lip.

the left foot were also excised, but the patient wished to keep the accessory digit and toe on the right side.

DISCUSSION

The rarity of median upper lip clefts is well recognised. Fogh-Andersen (1965) reported 15 cases in a survey of 3,988 facial cleft patients seen at one hospital over a 30-year period.

Median clefts may be associated with digital abnormalities (as in three of Fogh-Andersen's cases). There may, in addition, be gross systemic abnormalities and the patient may be mentally deficient. The digital defects may include clinodactyly and asymmetrical shortening with or without syndactyly, as in the Oral-facial-digital syndrome. Polydactyly is also seen with median clefts in the Median-cleft short rib polydactyly syndrome in which affected infants have major skeletal, renal and vascular abnormalities and die from gross respiratory tract malformations.

This case demonstrates a minor variation of the association between median clefts of the lip and digital abnormalities. The patient was of normal intelligence and the appearance of the hands was not strikingly abnormal. The extra digits were well formed and the anatomical proportions of the hand were little affected. Thurston (1909) described similar findings in a 20-year-old recruit for the Bengal police. He noted that the patient's younger brother had seven fingers on each hand, six left toes and a normal right foot. The cleft was similar to the cleft seen in our patient.

Burian's (1962) case had a more extensive median cleft, extending into the philtral region, bimanual hexadactyly and a double first toe on each foot. Direct comparison between all these cases is possible because photographs have been published with the reports.

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