

# Aspects of Plastic Surgery

## Cleft Lip and Palate—General

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*British Medical Journal*, 1974, 3, 107-108

### Introduction

Cleft lip and palate is one of the commonest of the severe congenital deformities and some degree of the deformity occurs in about one in 800 births. During the last 30 years there have been very considerable improvements in the overall results of treatment. Nevertheless, few of these have been due to advances in surgical techniques, which have changed remarkably little since the principles in this country were laid down by Kilner and Wardill in the 1920s. Instead, they have been brought about by the following:

The improved health of infants in general, due partly to improved social conditions and partly to developments in paediatric health services.

Improved methods of anaesthesia which make it possible for the surgeon to work at greater leisure and with more precision and which have a very low incidence of postoperative complications.

Improved follow-up facilities by multidisciplinary teams which make it possible for later-staged treatment to be carried out at optimum times.

A greater knowledge of, and improved treatment of, hearing defects in children. In particular, the use of the operating microscope has made it possible to preserve good hearing in patients who might previously have suffered severe hearing losses and consequently been at great disadvantage when their speech was developing.

### Embryology

Though recent investigations have suggested modifications and refinements, the classical hypothesis put forward by His still provides a satisfactory background for understanding the clinical deformities (see fig. 1).

During the fourth week of intrauterine life the maxillary processes appear below the developing orbits (fig. 1a) and grow forward to fuse by the sixth week with the frontonasal process (fig. 1b). Thus, the frontonasal process forms the columella, the central part of the lip—that is, the philtrum—and the pre-maxilla, which contains the incisor teeth. The maxillary process forms the lateral part of the lip and the dental arch as far forward and including the canine tooth.

Fusion of the processes is originally only by epithelium, but it is consolidated later by mesoderm, which invades the fusion line mainly from the maxillary process. This stage of development results in the formation of the lip, the alveolus, and the extreme anterior end of the hard palate, and it is now usual to refer to these parts as the lip and primary palate (fig. 1c). Failure of development at this stage could be due to failure of the processes to meet, but some workers believe that the most likely cause of deformity is failure of the mesodermal invasion. If mesodermal invasion does not occur, the line of epithelial fusion is unstable and breaks down, resulting in a cleft of the lip and

alveolus. Nevertheless, even in a complete cleft a thin band of tissue may sometimes be seen connecting the two sides. This is thought to represent the remains of the early epithelial fusion.

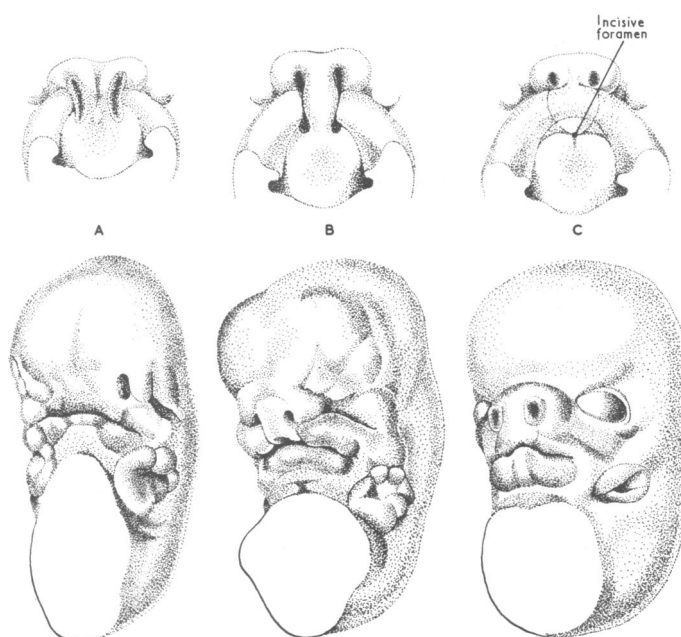


FIG. 1—Formation of the lip and primary palate as far back as the incisive foramen. The maxillary process can be seen growing forward and fusing with the frontonasal process and this union then becomes consolidated. The upper row shows the view from the palatal aspect and the lower row shows the external view at the same stages.

It was previously thought that the process of mesodermal invasion commenced at the level of the alveolus and spread backwards and forwards from this point. It now appears more likely that the process of invasion starts further back in the anterior part of the hard palate at a point which in the fully-formed palate is marked by the incisive foramen. The mesodermal invasion spreads from this point forward but may become arrested at any stage, so that a failure of this process results in a cleft which may extend backwards from the free edge of the lip to any point as far posteriorly as the incisive foramen.

The palate behind the position of the incisive foramen is formed at a later and distinctly separate stage by the outgrowth of palatal shelves from the maxillary processes. These appear during the sixth week and fuse with each other and with the developing septum in the midline during the eighth week. This fusion takes place from the incisive foramen backwards so that a cleft of the secondary palate may extend from the uvula to any point as far forward as the incisive foramen (fig. 2).

Failure of development may occur during the stage of lip and primary palate only, of the secondary palate only, or of both stages.

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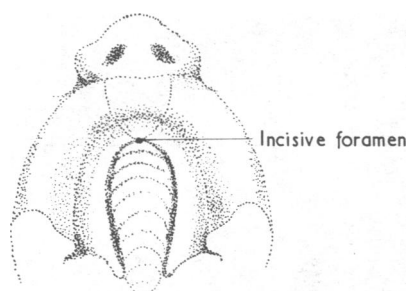


FIG. 2—This shows how the palatal shelves appear and join from the incisive foramen backwards.

### Anatomy and Classification

Partial or complete clefts of the lip and primary palate or the secondary palate may exist alone or in combination and may be unilateral (fig. 3) or bilateral (fig. 4). In the complete form a continuous cleft exists from the free edge of the lip to the posterior end of the soft palate. The cleft of the lip is always associated with distortion of the nostrils and of the nasal septum.

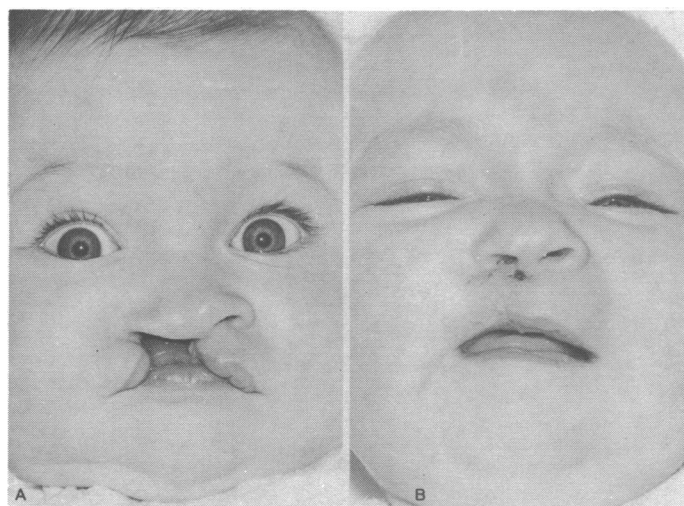


FIG. 3—Complete unilateral cleft of the lip and palate. The lip has been repaired by Millard's method.

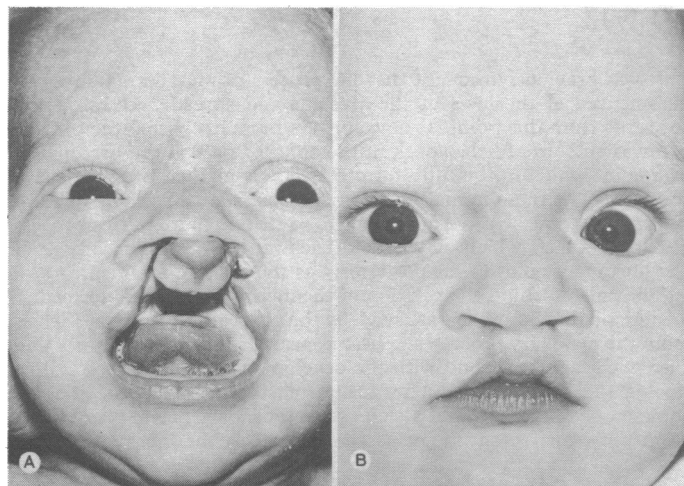


FIG. 4—Complete bilateral cleft of the lip and palate. The postoperative result shows that the columnella is still very short with pulling down of the tip of the nose and this will need correction at the age of 4½.

### Causation

It now seems certain that several factors must be operative before a cleft develops, some of them genetic and some of them existing in the environment of the developing embryo. From the genetic point of view three groups may be recognized:

(1) Complete unilateral or bilateral clefts of the lip and palate or incomplete manifestations of this basic pattern. In this group there is a Mendelian recessive factor, but in addition to the genetic factor, it seems certain that unknown environmental factors must be present before the deformity becomes manifest.

(2) Clefts of the secondary palate only, never involving the primary palate or lip, and with a strong family history. Deformities in this group are confined to females.

(3) Clefts of the secondary palate only, without involvement of the primary palate or lip, and without any family history of similar defect. These defects occur quite sporadically and are presumed to be due entirely to environmental effects on the developing embryo. Some babies in this group have severe underdevelopment of the mandible, which causes difficulty in feeding and respiration—the so-called Pierre-Robin syndrome. Strong evidence suggests that the deformity is caused by pressure in utero associated with the extreme flexion of the neck so that the chin is pressed on the sternum and the tongue is pressed forcibly into the roof of the mouth, thus interfering with the approximation of the palatal shelves.<sup>1</sup>

### Genetic Counselling

In group (3) there is no greater risk of further children of the same parents of children of a deformed child having a greater likelihood of developing clefts than in the general population. In group (2) it is very likely that any further female children or female children of the affected person will have clefts, and these patients should therefore be advised accordingly. In group (1), which is the commonest type of cleft palate deformity, very often no history of similar defects in close ancestors can be elicited and the chances of further children being affected are small. Even so, if a close relative of one or other parent is similarly affected, then the chances of further children being deformed are very greatly increased and are probably about one in 80. If there is a family history on the side of both parents then the dangers of further children being affected are very great indeed.

### Changes in Facial Growth after Birth

In addition to non-union of the parts there is also an actual lack of tissue, both soft tissue and bone, in the region of the cleft. We thus have both distortion and lack of tissue. In the past it has been difficult to know how much of the changes in facial growth after birth and after operative treatment were due to the initial deformity and how much to the effects of treatment. In general, it may be said that the operative repair creates forces which tend to improve the alignment of the underlying skeleton but that the deformity due to lack of bone will persist. So far as the upper dental arch is concerned, a properly executed operation creates forces which tend to improve the shape of the arch anteriorly. Nevertheless, further back in the region of the premolars a repair of the cleft creates unfavourable forces which tend to cause contraction of the arch. Under modern conditions this deformity is small and the benefits of operation far outweigh the disadvantages. The very bad deformities of the maxilla which have been seen in the past were almost certainly the result of bad surgery.

### Reference

- <sup>1</sup> Poswillo, D., and Roy, L. J., *British Journal of Surgery*, 1965, 52, 902.