HARE-LIP AND CLEFT PALATE IN DOUBLE-MONSTERS

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A large volume of literature is available on the biology of identical twins from the genetic standpoint. The diagnosis of monozygotic twins though based on a number of anatomical characters establishes only the great probability of the twins being identical as a fairly reliable inference. But in cases of conjoint twins or double-monsters we have unimpeachable evidence of the twins being split portions of the same embryonic disc and hence absolutely positive factual evidence of identity in genetic make-up. The material available for such study, *i.e.*, double-monsters, is very scarce, and hence the following observations are considered worthy of publication.

The occurrence of hare-lip and cleft palate in both components of a double-monster was noticed in a specimen dissected by one of us in 1928 (Specimen 1, Fig. 1). Since then we have been on the lookout for a similar condition to gather additional evidence. Observations have been made on three cases of double-monsters in which both components show the abnormality of hare-lip and/or cleft palate. In all of them the defect is symmetrical so that one component is a mirror image of the other.

Report of Cases

Two of the specimens are human fœtuses and the third is a double-headed kid. Diagrams 1, 2 and 3 show the appearance of the specimens.

Specimen 1.—The specimen is a human double-monster, female, thoracopagus tetrabrachius tetrapes. The union is extensive on one flank from the upper part of the thoracic region to umbilical region. Both faces show unilateral hare-lip and cleft palate, the right component on its left side and the left component on its right side, so that each is a mirror image of the other.

Specimen 2.—This is a human monster with single trunk and a fused double head showing two faces, diprosopus distomus Triophthalmus, diotus. Each face shows a bilateral hare-lip with cleft palate.

Specimen 3.—This is a double-headed male kid with cleft palate in both components. The palatal cleft in each is median with defective nasal septum which is a sickle-shaped ridge at the roof of the nasal cavity.

It is not necessary for the purpose of this paper to give a detailed description of the abnormality in the specimens as the significant fact is the symmetrical occurrence of the anomaly in both components and especially the mirror imaging of the unilateral anomaly in the first specimen.

DISCUSSION

Among the various etiological factors for the conditions, hare-lip and cleft palate, the following are usually cited.

1. Local factors like amniotic bands preventing the normal union of the processes constituting the face or non-union due to circulatory failure (Keith, 1940) or pressure caused by the lower jaw and tongue forcing out the processes forming the upper lip or palate in instances where the hyperflexed chin abutted against the chest or knee (Cryer, cited by Mead, 1946).

2. Injury to a part of the embryonic mozaic due to environmental factors at a critical stage in early development leading to defect or malformation of the part of the body derived therefrom (Stockhard, Ballantyne, cited by Willis, 1950; and Murphy, 1947).

3. Hereditary factor transmitted as a mendelian inheritance (Gruneberg, cited by Browne, 1950; Gates, 1946, and many others).

In individual cases where the presence of amniotic bands or local causes leading to pressure on or necrosis of embryonic parts have been definitely made out, it cannot be denied that such factors do have an etiological bearing. But in the instances presented here, showing symmetrical deformity of the two components, purely local factors could reasonably be eliminated.

There is ample and accumulating evidence concerning the correlation between defective development and anoxia, chemical poison, or bacterial or viral toxin present in the environment at critical stages of early embryonic development. If such factors are considered as being operative causes in the instances of double-monsters presented here, one would naturally expect a bilateral defect in each of the components of the double-monster because the facial processes that go to form the upper lip of the embryo appear synchronously and progress to their fusion simultaneously on the right and left side of the embryo. Such a hypothesis may explain the condition in

Specimens 2 and 3 but not the condition in Specimen 1. There is no evidence yet to assume that the stages of embryological development on the right and left halves of the upper lip are asynchronous and that one lateral half of the upper lip is constituted in advance, while the other half lags behind in development. The absence of a bilateral defect in both components in Specimen 1 would eliminate the possibility of toxic influence at a critical stage during development as a causative factor.

Further, the fact that the anomaly is subservient to the law of symmetry of the total embryonic primordium composed of both components, *i.e.*, the fact that one component is a mirror image of the other in unilateral asymmetry would lead us to infer that we have to deal here with an inherent tendency of the embryo to develop in a malformed manner. This tendency conduces to the production of an anomaly of a particular kind, but not necessarily focussed to any one particular antimeric side of the embryo. In its ultimate expression this tendency is seen to be conditioned by the compelling law of symmetry as applicable to the total embryonic area, as if an axial line for symmetry were passing through the conjoint portion of the double-monster. Such a tendency could only be a genetic factor. In such a case it could vary in its penetrance and expression from parent to offspring and even on the two sides of the individual. Being carried by the genes in the chromosomes its transmission from one generation to another would be in the manner of mendelian inheritance.

The symmetrical occurrence of unilateral hare-lip and cleft palate in a double-monster is an additional evidence supporting the genetic factor in the etiology of these conditions.

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Summary

Three cases of hare-lip and/or cleft palate occurring in both components in double-monsters in a symmetrical manner are recorded. One of the specimens shows unilateral hare-lip and cleft palate occurring on the near side of the two components, each mirror imaging the other in its individual asymmetry. This adduces additional evidence to the genetic factor in the etiology of these conditions.

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