9-1-1934

Wolf Snout And Other Anomalies In Monovular Twins : A Case report

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Reprinted from the FREEDMEN'S HOSPITAL BULLETIN, *September, 1934.*

"Wolf Snout" and Other Anomalies in Monovular Twins: A Case Report*

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History

In February, 1929, premature monovular male twins presenting developmental anomalies here described were delivered in Freedmen's Hospital†. An interval of 10 minutes separated the births. Both infants cried spontaneously. The first infant, Dee, weighed 4 lbs. 4 oz. and lived 25 hours, 15 minutes. The second infant, Dum, weighed 4 lbs. and lived 8 hours, 20 minutes. Autopsies were not performed and the causes of death were not definitely determined. The remains were transferred to the Anatomical Department of Howard University where they were recently studied.

The fetal membranes consisted of two amnia and a single chorion attached to a large placenta into which both cords were peripherally inserted. This finding and the facts that the infants were of the same sex, nearly the same size, and exhibited an external developmental defect of similar character and degree, were together considered strong evidence of uniovular origin.

The mother was an American Negro of 23 years. She had previously borne three children, all of whom were "deformed" and soon died, and had suffered one miscarriage. The nature of the "deformities" could not be learned. The probable duration of pregnancy in the case of the present twins was 8 lunar months. Other significant history was not available. The mother's Wassermann reaction, at first negative, became strongly positive after provocative stimulation.

Physical Findings

In size, as in weight, Dee slightly exceeded Dum. Although complete sets of dimensional measurements and organ weights were taken, the data are not tabulated because they do not show additional significant or reliable differences between the two infants.

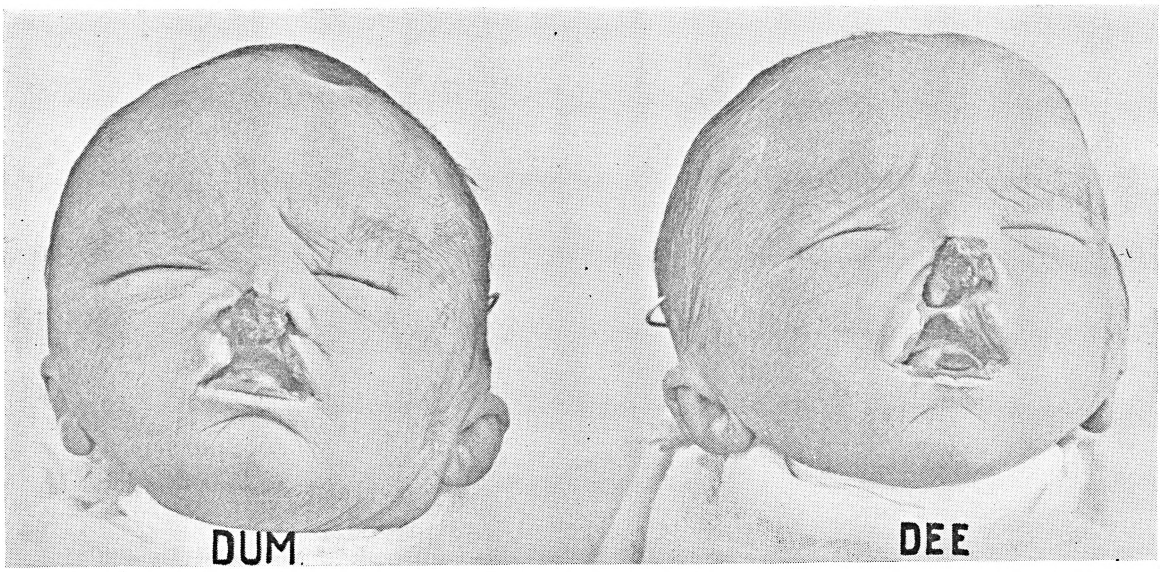

Fig. 1. Bilateral total cheilognathoschisis (wolf snout) with absence of the middle segment of the upper lip.

Face: Both specimens exhibited bilateral cheilo-gnatho-schisis, with absence of the middle segment of the upper lip, an exaggeration of the deformity commonly known as "wolf snout," Fig. 1. The median tubercle or premaxillary element protruded beyond the rest of the race. It was reduced in size and bore incomplete dental sacs which exposed partially calcified medial incisors. The element was not united to the maxillary palate behind, but was continuous above with an enlarged nasal septum. The septum itself was not completely joined to the palate but passed 3 mm in Dum and 2.5 mm in Dee posterior to its anterior border before uniting.

Lateral and adjacent to the dental sac of the left medial incisor in both individuals was a small area, outlined by a dark line, of about half the size of the medial sac and resembling

*Presented before the American Association of Physical Anthropologists meeting in New York City, May 9, 1934.

†Obst. No. 285, 1929; Births Nos. 239 and 245, 1929.

it in shape. This area was the outer surface of a prominence which appeared to be partially folded behind the central incisor. The shape and location of this prominence suggested that it represented the dental sac of the lateral incisor. Beside the right medial incisors there were no indications of laterals.

..The lip was totally absent over the premaxillary area. As a result the gum above the dental sacs was separated from the broad tip of the nose only by a narrow groove covered with skin. The portion of the palate formed by the lateral palatine processes was intact. Demonstrable reduction in size of the maxillae was not present.

Laterally, facial development approached the normal. The outer segments of the upper lip were correctly formed and the lateral three fourths of the vestibules of the nostrils were well modelled. The inner fourth of the nasal vestibules was also distinctly outlined but the outer and inner portions were interrupted by thinned notch-like areas. These were more marked in Dee than in Dum, and apparently were secondary defects due to stretching by the forward and slightly upward projection of the unrestrained premaxillary region.

The glabrous and villous portions of the mucous membrane of the lips were distinctly demarcated. The lateral segments of the upper lips appeared slightly hypertrophied in comparison with the lower lips. The natural borders of the lateral segments formed slightly obtuse angles with those bounding the gap, thus making it appear as though a slice had been removed from the upper lip. The skin and mucous membrance had united over the abnormal margin. This mucous membrane showed from below upward on the face, the glabrous portion, the pars cillosa and internal mucosa. Only the latter and skin were visible at the junction of the lips to the nasal vestibules. There was no secondary cutaneous anchorage of the lateral lip segments to a receded maxilla.

The tissue deficiency in the upper lips was negligible. The sum of the lengths of the external mucous membrance of the upper segments equalled the length of the lower lip in both infants. The tissue deficiency of the external nares was but slightly greater. The median tubercle had the appearance of having been pushed through the front of the face and so prevented proper fusion of the facial components.

It was clearly evident that the aborted components were the medial nasal processes and their derivatives the medial palatine processes. These elements were fused in the midline but had been symmetrically inhibited at their periphery. No lip had formed from the medial nasal processes and there had not been sufficient growth to accommodate properly the lateral incisors in the median palatine processes. Consequently the premaxillary element had remained widely separate from the palate, a hiatus left between the portions of the lip formed from the maxillary processes, and the vestibules of the nostrils secondarily divided by the forward stretching of the independent central process.

Great Vessels: The patterns of the great venous channels which converge to the heart were of the common type and similar in the twins. The great arterial trunks presented unusual anomalies.

In both infants, the common carotid arteries arose together and the subclavian arteries arose together, Fig. 2. The common carotids passed upward in front of the great veins of the neck, and the right subclavians passed behind the oesophagus. There was no innominate artery in either infant. The twins differed in that the arch of the aorta, that segment extending between the origins of the carotid and subclavian arteries, was absent in Dum. The figures show clearly, however, that this element was reduced even in Dee, an observation which alone would not arouse much interest.

Heart: The foramina ovalia showed similar expected patency. There was no defect in the interventricular septum of either heart, the aortic and pulmonary trunks thus being completely separate at their origin. The coronary arteries in both hearts were derived from the aortic root.

Kidneys: The kidneys and their vessels in Dum showed no abnormal features. The left kidney of Dee was normal. The right kidney of Dee, Fig. 4, was congenitally displaced and lay over the bifurcation of the aorta. The hilus was directed anteriorly, having retained its primitive position. The blood supply was from a renal artery given off from the aorta just above the inferior mesenteric artery. No adventitious vessels were noted. A single renal vein which emerged from the hilus at the same horizontal level at which the artery entered, emptied into the left renal vein just lateral to the left side of the aorta. The ureter proceeded directly downward from beneath the vessels. The right kidney bore a Y-shaped impression of the bifurcation of the aorta and inferior vena cava posteriorly and an impression of the left ureter laterally.

Lungs: The right lung of Dee had four lobes. The supernumerary unit was an apical division of the upper lobe.

Testes: Both testes of Dum and the left testis of Dee had descended into the scrotum. The right testis of Dee lay just before the internal abdominal ring.

Additional: To detect further resemblances or dissimilarities between the twins, a few selected features were compared.

The occipital hair whorls had the same general pattern but differed distinctly in certain details‡.

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‡The photographic records are available in our files.

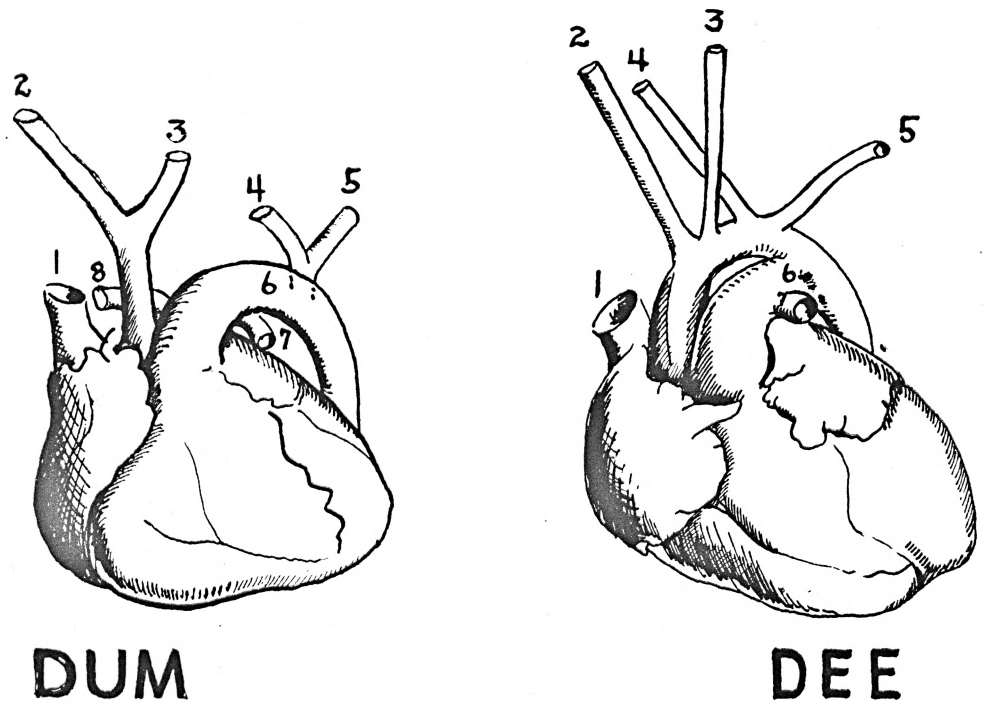

Fig. 2. Anomalous origins of common carotid and right subclavian arteries. Absence of aortic arch (Dum). 1. Sup. vena cava 2. R. com. carotid 3. L. com. carotid 4. R. subclavian 5. L. subclavian 6. Ductus arteriosus 7. L. pulmon,art. 8. R. pulmon, art.

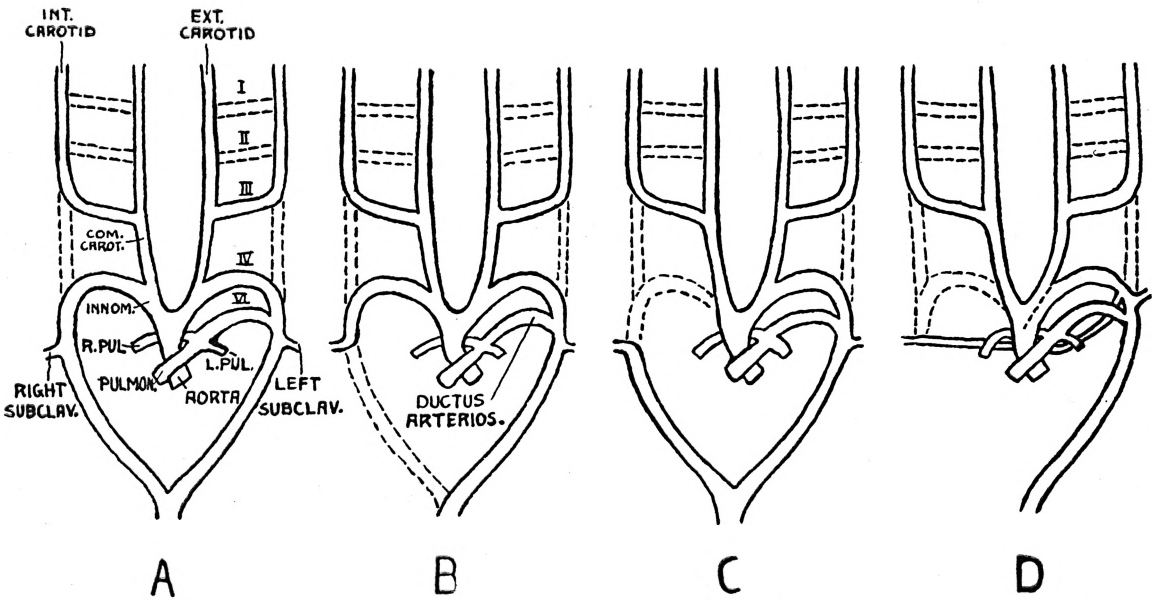

Fig. 3. Diagram to show embryonic origin of anomalies in Figure 2. A. Primitive pattern with left and right aortic arches complete. B. Usual mode of formation of right subclavian from proximal portion of right aortic arch. C. Anomalous formation of right subclavian from distal portion of right aortic arch. D. Upward shift of right subclavian on aortic arch to position near left subclavian. Also, mode of formation of single origin for both common carotids.

The configuration of the four external ears showed only individual variation‡.

The femoral arterial complexes were largely similar. The lateral circumflex artery arose from the profunda on both sides in Dum and Dee. The medial circumflex was derived from the femoral on both sides in Dum and on the right in Dee. This vessel arose from the profunda on the left in Dee. In Dum there was on the right an accessory vessel which arose from the femoral and accompanied the descending branch of the lateral circumflex.

The lungs, livers, spleens, and thymi of the two infants differed perceptibly in form as well as in size.

Discussion

Face: The occurrence of clefts in the lip or palate in each individual of twins is very rare. Lévy[1] describes a case and mentions those of Appert and Cargile. Reports of the latter authors were not available. Lévy's case, like ours, was of premature male monovular twins of eight months, with separate amnia but sharing one placenta. Each of the infants had right unilateral harelip and one had also a palatine fissure. Both left the hospital in good condition on the twenty-fifth day. The mother's Wassermann was negative.

No additional cases are cited in five statistical surveys[2, 3, 4, 5, 6]. In children of the same parents Haug[3] found two individuals affected thirty-two times, three individuals once, and four individuals three times. Three of the cases in Peron's[5] series were in twins of whom the other member was normal. The sex was the same in one of these pair. No case was encountered in which the anomalies exactly corresponded with those described here. The hereditary occurrence of clefts of the lip and palate is placed between 15 and 20 per cent of cases by various authors[4]. Because of a positive history in 35 per cent of his cases Peron[5] concluded that lues was an important contributory factor. Davis[4] discounted low mentality and social status as predisposing causes. Similar deformity in monovular twins affecting other parts of the body is not uncommon[1].

Cadenat[7] offers an explanation of the pathogenesis of the defects in our specimens. He states that in the earliest stages of development the face and medial nasal processes are supplied by the cerebral branches of the internal carotid. Later with the invasion of the musculature by the facial nerve and artery there is a change in blood supply to the external carotid. At the same time the internal carotid withdraws to become specialized for the supply of the enormously developing brain. He remarks that Tandler has shown in a fetus with bilateral total cleft that the primitive septal branch from the internal carotid is well developed and supplies the median tubercle. Since it has become established that embryonic events must occur at specific times or the regions involved will be abnormal or retarded, it may be conceived that if for any reason the medial nasal processes should not fuse with the maxillarys in those very busy weeks (5th through 8th) of embryonic life when so many organs are passing through critical stages, the median tubercle will thereafter suffer a deficiency in blood supply and remain permanently dwarfed. It is difficult, nevertheless, to admit of blood vessels withdrawing from a region as long as there is anything for them to supply.

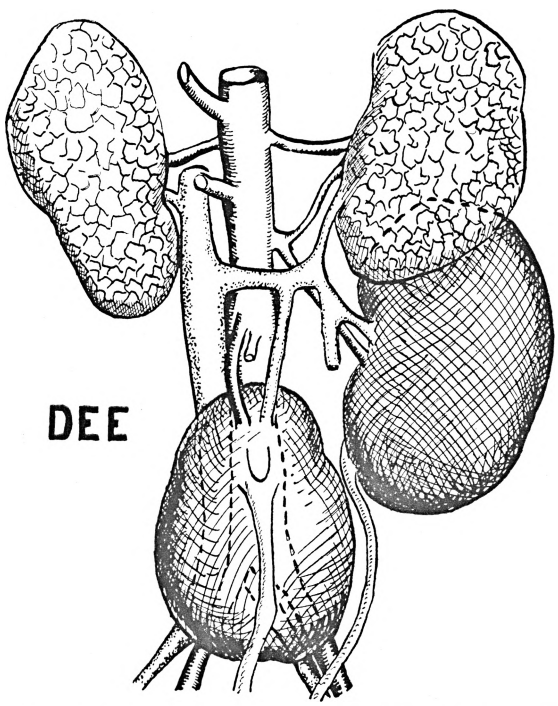

Fig. 4. Congenital displacement of right kidney, with anterior position of hilus, and right renal vein emptying into left renal vein.

Veau[8, 9] affirms that although the median tubercle forms the middle segment of the upper lip, the latter derives its muscle, blood and nerve supply from the maxillary process. By elimination, skin and mucosa only are left to be formed from the median process. With a restricted blood supply it is possible that this segment might become inhibited altogether as in our specimens. Veau[8] seems convinced, however, that the median tubercle does contribute perhaps all of the premaxillary

‡The photographic records are available in our files.

bone, including the lateral incisors. He includes a review of the controversy on the subject§. It is still being investigated.

The pathogenesis of these defects thus remains obscure. Their occurrence in families and in monovular twins seems to indicate genetic factors. The presence of a graded series of the malformations in a litter of puppies[10] suggests the possibility of Mendelian inheritance, since multiple ova are released at an ovulation in dogs. Keith was unable to produce the defects in lion cubs by altering the nutritive conditions[10].

Great Vessels: Normally the right subclavian artery is formed as far as its segmental unit, by the proximal portion of the right aortic arch, and the distal portion of this arch disappears, Fig. 3b. When the proximal portion of the right arch disappears, the distal portion contributes to the right subclavian and thus gives the latter an anomalous origin from the (left) aortic arch, Fig. 3c. In such cases, obviously, there can be no innominate artery. Thomson found such an origin for the right subclavian in 1 per cent of 500 cases‖.

Due to subsequent developmental changes the right subclavian may migrate any distance up the aorta. In our cases the artery has attained the level of its fellow of the left side. The comprehensive monograph of Holzapfel[11] and the case report of Cobey[12] furnish details of the embryonic mechanics of this process.

It is apparent that a high fusion of the primitive ventral aortae (common carotids) or a low origin of the fourth left aortic arch (arch of the aorta) would produce a common origin of the two common carotids, such as occurs in Dum and is approximated in Dee. In Dee the aortic arch is reduced and in Dum it is absent. In the latter the functional aortic arch is formed from the pulmonary trunk continued as the ductus arteriosus or embryologically the sixth instead of the fourth primitive aortic arch.

The primitive aortic arch system repeated in human ontogeny is more complicated than the primitive cardinal venous system about the heart. Consequently variations and anomalies in the great arterial vessels are more frequent than in the great veins though anomalies in the latter are not uncommon.

Kidneys: Although anomalies of the renal vessels are common, congenital displacement of the kidneys is rare. In a review of 20,000 autopsies Guizetti and Pariset[13] found 18 cases. Thomas[14] found 5 cases in 1,800 autopsies. The latter author found the condition more frequently on the left in the proportion of 65 to 36 in 101 cases.

§The position of the lateral incisor in cases of total harelip shows considerable variation, occurring both in the medial nasal and maxillary processes. Keith[10] has seen this member straddling the cleft. It is probable that the dental lamina as a dermal derivative possesses some developmental independence of the nasal and maxillary processes.

‖Quoted by Cobey.

Because the region of the kidneys and inferior vena cava, like that of the heart, is the scene of rapid recapitulatory and tachygenetic changes during embryonic life, it is especially liable to developmental, accidents, such as the persistence of early stages. As a result, a congenitally displaced kidney is nearly always low. In our case it is both low and has failed to assume a lateral position and rotate inward. Specimens T.15 and T.87 from the Western Reserve University teratological collection which have been described by Huffman[15], illustrate further the conditions involved in congenital renal displacements.

Additional: It is well recognized that uniovular or identical twins are not identical. The differences between the individuals may be marked as in parasite-autosite monsters or they may be slight. The degree of resemblance of monovular twins, especially in the mental sphere is still under investigation.

In twins of defective constitution such as our specimens it is to be expected that dissimilarities would be found in the regions most liable to developmental defects, where in our specimens defects did occur and dissimilarities were found. It is impossible to estimate what measure of the defects described was due to hereditary factors and what to environmental influences. The work of Mall[16] and Stockard[17] has shown the potency of environmental conditions[8]acting at critical embryonic periods in producing malformations in lower vertebrates. It is certain, too, that even in the uterus litter mates are not affected by precisely the same environmental influences.

Summary

1. In newborn male monovular twins of probable luetic parentage, bilateral total cheilognathoschisis and anomalies of the great vessels of the heart, a kidney, a lung, and a testis are described.

2. In selected features resemblances and dissimilarities between the two infants are indicated.

3. The findings are discussed in relation to their causation, hereditary and environmental influences.

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N. B.—The writer is grateful to Dr. N. W. Ingalls for critically reviewing the contents of this article.