AGLOSSIA CONGENITA WITH BONY FUSION OF THE JAWS

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AGLOSSIA congenita is an extremely unusual and rare condition. The first case which appeared in the literature was that described by De Jussieu in 1718, who reported this form of abnormality in a Portuguese girl aged 15 years who had merely a small, wart-like, mobile muscular swelling in place of the normal tongue. Since that time several examples of the disorder have been reported, and reference has been made to co-existing defects of the soft and hard palates. In some instances, too, severe defects of the hands and feet have been observed in individuals with a rudimentary tongue structure.

Fulford (1956) has published a comprehensive account of aglossia congenita referring to cases recorded by De Jussieu, Rosenthal, Spiller, Kettner, Watkins, Sinclair and McKay and Farrington and Fitzwilliam. This author's survey deals particularly with the embryological aspects of the deformity.

A case of aglossia congenita with bony fusion of the jaws was described by Petterson in 1961. It concerned a male infant who was unable to open his mouth at birth. The second disability was found to be due to anterior fusion between the maxilla and mandible. The child's lips were normal but the chin was recessive in type. A full description of the author's operative procedure is given and two oral anomalies confirmed namely, a cleft of the soft palate and a mucosal wall connecting the palate and the floor of the mouth. In addition malformations of the hands with absence or poor development of the nails was noted. Swallowing and speech difficulties are often experienced by individuals suffering from this congenital affliction.

Embryology. The complex process of tongue development must have been interrupted at an early stage in affected persons, and in Fulford's opinion failure of tongue development is most commonly due to agenesis of the first mandibular arch. The tongue rudiments in the cases reported were all mobile aud contained innervated muscle. Fulford further contends that the occipital somites and their nerves migrate normally, but, because of the previous agenesis of the first arch cannot form the normal musculature. The narrow face with receding chin is due to a hypoplasia of the mandible and a narrow mandibular arch. The intra-oral band between the palate and the floor of the mouth probably represents the remnants of the bucco-pharyngeal membrane. The association between aglossia congenita and deformity of the limbs is significant but difficult to explain.

CASE HISTORY

Baby C. was born at the Birmingham Maternity Hospital on 6.4.60. This was a normal delivery to a gravida 7 mother. The child was cyanosed for 10-15 minutes at birth and the only air entry was via the left nostril. The jaws were found to be fused and the mouth virtually imperforate, and because of this the infant was transferred to Birmingham Children's Hospital.

On first examination, the baby of Jamaican descent, showed a perfectly formed face and lips (Fig. 1), but the jaws were found to be fused when the lips were parted. It was not possible to pass a probe into the oral cavity. Other deformities noted at this stage were syndactyly of the left hand, gross failure of development of the right hand and a congenital amputation of the left foot.

The radiological examination of the jaws was reported on as follows: 'The lower jaw appears smaller than usual. The upper jaw is also hypoplastic particularly on the left, where the left orbit looks larger than the right. The nasal septum is deviated to the right, and a hard palate cannot be identified. Anteriorly there appears to be a fusion of the upper and lower alveolar margins' (Fig. 2).

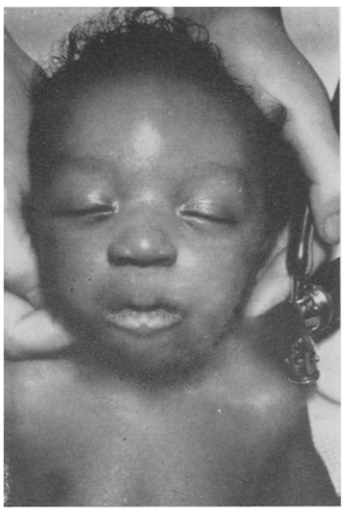

FIG. 1

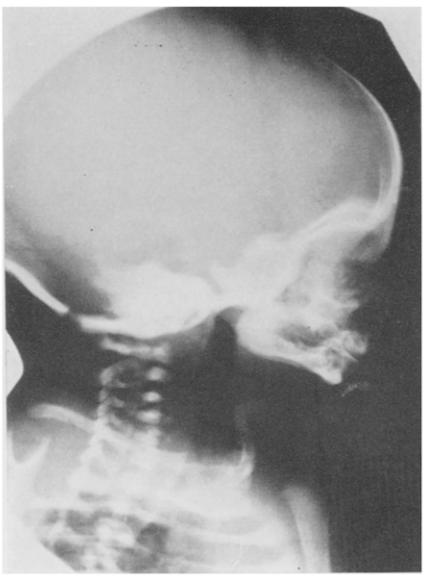

FIG. 2

FIG. 1.—Photograph of baby showing normal formation of face and lips and with a tracheostomy tube inserted before jaw operation.

FIG. 2.—Lateral view of skull and jaws demonstrating anterior fusion between maxilla and mandible

It was decided that in view of the otherwise robust nature of the infant, an operation to open the jaws should be carried out. A preliminary tracheostomy was performed, and a general anaesthetic was then administered by this route. A membranous curtain was found between the floor of the mouth and the upper jaw posteriorly and this was incised revealing an area of bony fusion about 2 cm. wide between the premaxilla and the mandible. This osseous mass was divided by chisels and it was then possible to open the jaws fully. The jaws were small, the mandible showing marked micrognathia.

At this stage the minute tongue was discovered which probably comprised only the posterior third and was firmly bound down by a membrane to the floor of the mouth. The organ was freed and a suture inserted to draw it forward and provide some control. The soft palate and half the hard palate (posterior part) were seen to be widely cleft.

Following the operation and after an initial period of respiratory difficulty the child thrived. The tracheostomy tube was removed at the end of four weeks from the date of operation.

The child was re-admitted in February 1961 for re-assessment at which time the general condition was only fair and a loose cough was present. The range of jaw movement was limited, and it was decided not to proceed with repair of the palate or treatment of the amputated parts until he was older and stronger.

Subsequent to this examination, the family moved northwards where he was transferred to one of Dr. Barnardo's Homes, since which time follow-up has been discontinued.

DISCUSSION

This case appears to be a severe example of the rare anomaly of aglossia congenita with bony fusion of the jaws and persistent buccopharyngeal membrane. Other defects include cleft of the soft palate and partial cleft of the hard palate, together with syndactyly and congenital amputations. The social implications of family background and movement around the country has magnified the difficulties of treatment and management. Enquiries have failed to confirm that thalidomide was prescribed for the mother during the period of this pregnancy, and therefore the case must be viewed as a true example of this particularly serious and unfortunate congenital abnormality.

SUMMARY

The case of a Jamaican baby has been described who was born with an imperforate mouth due to the complex and very rare congenital disorder of aglossia congenita. Other congenital manifestations included bony fusion of the jaws, clefts of the hard and soft palates, congenital amputations of the right hand and left foot together with syndactyly of the left hand.

REFERENCES

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