AMBLYOPIA AND DISORDERS OF OCULAR MOTILITY IN CRANIOSYNOSTOSIS

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Abstract
In a further study of 39 cases of craniosynostosis, the timing of the Tessier procedure and of the squint surgery are discussed with reference to the incidence of amblyopia and optic atrophy. The effects of the Tessier procedure on the horizontal deviation and particularly on the vertical deviation and the levator function are also discussed.

Key Words
Craniosynostosis, Tessier procedure, amblyopia, optic atrophy, squint, ptosis, hypermetropia.

The term craniosynostosis is applied to a group of conditions in early childhood in which one or more of the cranial sutures fuse prematurely. As the brain expands compensatory growth occurs across the uninvolved sutures leading to various skull deformities. This compensatory growth may be insufficient to prevent a rise in intracranial pressure which is liable to lead to papilloedema, optic atrophy and bony changes around the orbit. The cribiform plate is displaced downwards by the raised intracranial pressure so that the orbital roof becomes more vertical and this leads to a shallowness of the orbits and a relative proptosis of the globes. This is enhanced when the maxilla is underdeveloped, as in Crouzon's disease and in Apert's syndrome. Hypertelorism is produced by an expansion of the ethmoids in temporal directions[1].

These concepts dictate a policy of advising early neurosurgical decompression in all cases of craniosynostosis which show signs of a raised intracranial pressure in order to try and prevent the development of optic atrophy and to limit the progress of secondary bony changes. The deformities of the craniosynostoses contribute to the lack of the development of binocular function and to the occurrence of abnormal ocular movements in many cases, but they may be ameliorated by the major craniofacial procedures described by Tessier[2,3]. If this operation is performed sufficiently early with subsequent squint surgery when appropriate, there should be a chance of achieving the development of some form of binocular function even if this is of an anomalous type, but in many cases this is an unrealistic ideal, as discussed later.

The purpose of this study is to establish the incidence of amblyopia in children with craniosynostosis and to what extent the amblyopia might have been prevented by early craniofacial and squint surgery, and also to assess the results of surgical treatment. The general features of the craniosynostoses have been discussed in a previous communication[4].

OPTIC ATROPHY AND AMBLYOPIA

This paper is concerned with 39 children who have been assessed at the Hospital for Sick Children, Great Ormond Street, over the past 3 years; 15 of the 39 cases were subjected to the Tessier procedure. 10 cases had optic atrophy with a corrected visual acuity which varied between 6/12 and perception of light, and 7 cases had amblyopia without optic atrophy so that in these cases the defective vision was simply the result of a squint, but amblyopia can also occur in the presence of optic atrophy so that the defect in the vision is exaggerated by the amblyopia.

The intercanthal distance in the cases showing amblyopia varied between 34 mm. and 60 mm., but there were only 3 cases in which the intercanthal distances were asymmetrical with the amblyopic eye on the side with the greater

displacement. It follows that an asymmetrical intercanthal distance is only a minor factor in the development of the amblyopia, with other precipitating factors such as a refractive error (particularly astigmatism or anisometropia) or a partial degree of optic atrophy, as more important factors, quite apart from some form of squint which causes a strabismic amblyopia.

STRABISMUS

Strabismus was found in all but 4 of the 39 cases, and in general terms there is a greater preponderance of an exodeviation in hypertelorism, a greater preponderance of an esodeviation in Apert's syndrome, and a more or less equal occurrence of exodeviation and esodeviation in Crouzon's disease (Fig. 1). In all the 35 cases showing squint, there was an associated V phenomenon with an overaction of the inferior obliques which is a characteristic feature of the V phenomenon.

	NAD	ESO	ESO + vertical	EXO	EXO + vertical	vertical
Crouzons		4		3		
Crouzons + Hypertelorism				1		
Aperts		7	1	1		1
Aperts + Hypertelorism					1	
Hypertelorism	3	2		8		
Plagiocephaly + Hypertelorism	1		4		2	

1 case = unilateral complete tarsorrhaphy

Figure 1. Nature of squint in this series of craniosynostosis.

REFRACTIVE ERRORS

30 cases in the series had a significant refractive error of 2 dioptres or more of hypermetropia or myopia, or of one dioptre or more of astigmatism in one or both eyes; the astigmatism of the less good eye varied between ±0.75 and ±5.00 dioptres. The increased incidence of hypermetopia and hypermetropic astigmatism in this series (Fig. 2) is possibly associated with the shallowness of the orbit, which is a characteristic feature of craniosynostosis.

REFRACTIVE ERRORS

Hypermetropia				10
Myopia				2
Astigmatism	– Hypermetropic	14	}	
	Myopic	2	}	
	Mixed	1	}	18
	Myopic one eye Hypermetropic one eye	1	}	

Figure 2. Types of refractive errors in this series of craniosynostosis.

RESULTS OF CRANIOFACIAL SURGERY ON STRABISMUS

15 of the 39 cases had craniofacial surgery (the Tessier procedure) and 14 of these cases had an associated squint. In 7 cases of Crouzon's disease or Apert's syndrome the frontal bone and the maxilla were advanced with a reduction in the proptosis. The craniofacial surgery increased the inferior oblique overactions from a vertical point of view by up to 20 prism dioptres, but reduced the torsional influence of the muscles. In 8 cases of hypertelorism the craniofacial surgery caused a reduction in the hypertelorism with the persistence of a divergent squint in only 3 cases, and otherwise with the development of a convergent squint in a case with a previous exotropia or with an increase in the esotropia which was present before the craniofacial surgery (Fig. 3).

Pre-Operative Deviation	Post-Operative Deviation	Number of Cases
NAD	ESO	1
EXO	ESO	2
EXO	EXO	3
ESO	increased ESO	2

Figure 3. Influence of craniofacial operation on squint in hypertelorism.

RESULTS OF CRANIOFACIAL SURGERY ON PTOSIS

Ptosis may be present in some cases preoperatively, but when it develops post-operatively it may be the result of post-operative oedema, a disinsertion of the aponeurosis of the levator tendon, a retroposition of the globe, a loss of support of Whitnall's ligament (the superior transverse ligament) due to a mobilisation of the periosteum, or damage to the tendon or its nerve supply. It is evident, however, that although 75% of cases after craniofacial surgery show some degree of ptosis (as measured by the opening of the palpebral apertures), there is only a relatively slight degree of levator weakness when a detailed assessment is made of the levator function of the upper eyelid so that an actual involvement of the levator tendon or its nerve supply is relatively rare.

RESULTS OF SQUINT SURGERY

Squint surgery was carried out in 11 cases, including 7 cases following craniofacial surgery. The results show that only one case achieved a normal form of binocular function, with only a latent type of deviation. In the other cases there was a small residual horizontal deviation accom-

panied sometimes by a small residual vertical deviation, and in some cases 2 or even more operations were necessary to achieve a satisfactory cosmetic result. A small residual deviation, however, is consistent with the achievement of some form of anomalous peripheral fusion.

DISCUSSION

Only 24% of the patients in this series had optic atrophy so that theoretically it should have been possible to prevent defective vision in the other cases provided craniofacial and squint surgery were carried out sufficiently early. There is, however, a disadvantage in early surgical treatment of the squint because there tends to be a greater degree of relapse than when the surgical treatment is carried out in the older child. This is a contradiction to the usual management of a squint in childhood, when early surgical treatment enhances the possibilities of achieving a binocular result (even although this may be of an anomalous type), provided appropriate measures have been adopted pre-operatively to deal with any amblyopia of the squinting eye and also with any anomaly of retinal fixation. This difference in approach is related largely to the change in the character of the squint which follows craniofacial surgery, particularly when there is a significant increase in the vertical deviation. This tends to occur because an advancement of the frontal bone alters the position of the trochlea with a consequent change in the action of the superior oblique (Fig. 4), and an advancement of the maxillary bone alters the origin of the inferior oblique with a consequent change in its action in a similar way. These changes in the actions of the oblique muscles consist of a reduction in their torsional actions but with an increase in their vertical actions because of a change in the directions of the lines of pull. It must be assumed that the increase in the effectiveness of the vertical actions of the oblique muscles is greater for the inferior obliques than the superior obliques to account for the persistence of the V phenomenon, perhaps because an alteration in the action of each superior oblique in the region of the trochlea is a more or less inevitable outcome of the Tessier procedure[5].

The role of the superior obliques in the production of the V phenomenon in the craniosynostoses is defined more clearly in cases which have not been subjected to the Tessier procedure. For example, in hypertelorism there is a displacement laterally of each orbit, and this causes a decrease in

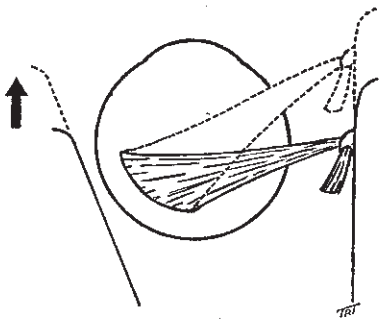

Figure 4. Change in action of superior oblique after craniofacial surgery.

the effectiveness of each superior oblique as a depressor of the eye because of an increase in the angle of pull of the muscle with the vertical meridian[5] (Fig. 5).

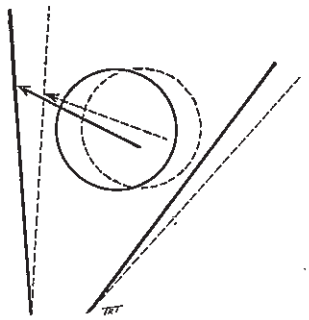

Figure 5. Change in line of pull of superior oblique in hypertelorism.
Continuous line represents orbit after lateral displacement and abnormal line of pull. (Drawing by Mr T. T. Tarrant, Institute of Ophthalmology)

A different view has been expressed[6] with an overaction of the inferior obliques as the primary event in the V phenomenon in oxycephaly and Crouzon's disease. This follows the shallowness of the orbit in an anteroposterior direction which results in a diminished role of the inferior transverse ligament of Lockwood (suspensory ligament) in supporting the eyeball so that there is an enhancement of the action of the inferior oblique and inferior rectus muscles which are closely associated with the inferior transverse ligament and which attempt to compensate for the diminished action of the ligament.

A further view has been put forward which suggests that the overaction of the inferior obliques in the V phenomenon in craniosynostosis is the result essentially of some form of muscular dystrophy[7], but this would appear to be a limited explanation in the majority of cases.

It is evident that there is some uncertainty about the cause of the oblique muscle imbalance which is a characteristic feature in craniosynostosis before and after craniofacial surgery. It is also evident that it is difficult to correct this imbalance by squint surgery, and this is complicated further by the change in position of the eyes which follows craniofacial surgery with an alteration in the horizontal deviation (a decrease in an exotropia or an increase in an esotropia), and an increase in the vertical deviation.

In general terms the management of such cases should consist of an early correction of any refractive error by spectacle lenses, the relief of amblyopia as far as possible by occlusion, and a delay in craniofacial and squint surgery until it is considered that the most appropriate time has been reached to achieve the best overall result.

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