AN ATYPICAL RESPONSE TO OCCLUSION OF A CONGENITAL ESOTROPE

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Abstract
Congenital esotropia of large degree generally has a poor prognosis for functional cure, unless early surgical intervention is undertaken.

Well supervised, pre-operative orthoptics is most important and may produce an unusual response. The congenital case studied was a large angle of deviation, which reduced dramatically following many months of orthoptics, beginning at the age of eight months.

Key Words
Congenital, esotropia, abduction, occlusion, binocular-function.

Introduction

Since 1777 partial occlusion of a sound eye had been suggested and since 1843 has been the orthodox treatment of strabismus when amblyopia of the strabismic eye is evident and was also recommended to "make the squinting eye straight."[1]

In recent times researchers in neuro-physiology, Hubel and Wiesel, have established experimentally with new born kittens, that when binocular pathway stimulation and binocular development is disrupted in early life, there is an anatomical deterioration of the binocular pathways in cells of the lateral geniculate body but no evidence of atrophy of the striate cells in the occipital cortex. [2,3,4] These findings would apply to both onset of strabismus and/or occlusion, as the disruptive factor. Hubel and Wiesel[2] state that "profoundly significant is the early disruption of the binocular pathways and rendering impossible any opportunity for fusion development. These changes seem to become irreversible after three months of monocular deprivation" in kittens. Their most recent findings state monocular deprivation is most devastating from the 4th to the 6th week of life in kittens.

If these findings can be related to human infants at any time, the chances of a congenital strabismus ever gaining functional bincular vision would seem remote.

Review of Literature regarding Congenital Esotropia

1. Worth 1908: – congenital esotropia is due to deficiency in a central fusion centre.[2]
2. Chavasse 1939:– binocular vision is achieved by conditioned reflexes and thus fusion is not an innate faculty and any obstacles to fusion development could be overcome if the deviation is eliminated before 2 years of age.[2]
3. Duke-Elder and Lyle:– little or no proof that functional cure was possible with early surgery.[1]
4. Costenbader 1952:– early surgery during the first 6 to 11 months of life, leaving a residual angle of 10 degrees or less, does result in functional cure. [2,5]
5. Berke 1958:– poor functional results with surgery after 2 years of age. [2]
6. Taylor 1963:– **adequate** surgery at an early age, i.e. 10 degrees or less laterally and 5 degrees or less vertically residual angle results in stereopsis,[6] and in 1967 poor results after 2 years of age.[7]
7. Costenbader and Parkes 1966:– early surgery resulted in 44.5% with gross stereopsis.[2]
8. von Noorden 1971:– 46% functional cure with adequate surgery up to 2 years of age.[8]
9. Taylor:– diagnosis of congenital strabismus cannot be made unless noticed prior to 6

months of age.[2]

10. Hubel and Wiesel:– by electro-medical testing of new-born kittens deduced the binocular pathways are innately determined and fusion faculty is anatomically present at birth.[4]
11. Duane in 1978 stated that "the management of congenital esotropia essentially involves surgically straightening the eyes".[9]
12. Walsh and Hoyt in 1969 stated "in most cases of convergent strabismus, any spontaneous straightening is associated with increasing amblyopia in the squinting eye."[10]
13. Dunlop and Dunlop (1978) found from an extensive survey of congenital squint only 4% gained "fully functional binocular vision"[12]
14. Selm (1974) reviewed 286 cases of congenital and infantile squints (being under 1 year of age) and found that refractive errors, particularly hypermetropia, are an insignificant causative factor in heterotropia of such early onset.[8]
15. Gillan (1945) analysed a series of squints treated with orthoptics alone and stated that 39% became straight or "nearly straight"[13]. It was not stated what treatment was given nor whether the deviations were intermittent or constant, congenital or acquired.
16. Guibor found one non-accommodative 20 degree squint which straightened with orthoptics alone. Again it is uncertain if this case was congenital, constant or intermittent.[14]
17. Costenbader (1958) like Chavasse stated "if vision is adequate in each eye, and binocular alignment is maintained during the first 6 years of life, fusion will often develop spontaneously."[15]
18. Marshall Parkes (1968) concludes that the younger the patient is at the time the congenital strabismus is straightened, the greater the success in the development of binocular single vision.[16]
19. Taylor (1963) concluded that early surgery for congenital strabismus should be carried out from 6 to 12 months of age, to enable the development of fusion and a functional cure.[6]
20. Duckman and Flax, optometrists, (1978) quote 76% overall functional and cosmetic cure by optometric orthoptic treatment of strabismus.[17] Perhaps much of the data tabulated involved intermittent and/or acquired strabismus, as there was no reference to congenital strabismus.
21. Forrest, an optometrist, (1977), cited a single case report of a congenital esotrope who suffered considerable birth trauma and presented at the age of 8 months with a 30 degree squint. With part-time occlusion and extensive "creeping and crawling therapy" and the "discouragement of walking", the angle of deviation finally reduced to 10 degrees with the use of binasal occlusion at the age of 2 years. He concluded that "there is little doubt that the forced lateral excursions had a positive affect on the results".[18] Possibly Forrest's case fits into the Blind-spot syndrome and this made a favourable situation for reduction of angle, once any suppression scotoma was eliminated. Alternatively, possibly it was a VI nerve palsy following birth trauma which recovered.

Case History

I have worked recently with a male infant with evidence of a large congenital constant right esotropia of 58 prism dioptres. Pregnancy proceeded normally and at birth the umbilical cord was around the infant's neck but presented no real problems. There is one cousin with a congenital esotropia. The child was first examined by an ophthalmologist under atropine at the age of 7 months, at which time there was no significant refractive error and fundi and media were normal. Orthoptic management was begun at 8 months of age and consisted of direct and constant occlusion which was not fully effective until 12 months of age and was supervised at regular monthly reviews. On presentation bilateral abduction was poor as is often found with congenital esotropia. Lyle and Wybar[11] suggest the possibility of stretching of the VIth cerebral abducens nerves in the process of moulding the head during birth as a cause. In spite of total and almost constant occlusion there was great reluctance to maintain fixation of the squinting eye. Easy fixation of the right eye was achieved at 17 months of age. I have suspected for some time that infants, in which reversal amblyopia occurs with constant occlusion therapy, have some form of bilateral foveal dysfunction or instability. Although there is no real evidence to support this at present, the incidence of bilateral eccentric fixation with bilateral sub-standard visual acuity which never reaches the now accepted foveal standard of 6/5 is surprisingly higher than one might have expected. At each monthly review a careful evaluation of clinical fixation was made. Much time was spent gauging any improvement of abduction and when able the Catford Drum, visual acuity was regularly assessed. Initially, the recorded visually acuity at 16 months was 6/12 R&L and

now at the age of 4 years the acuity is R.6/12 and L.6/9 with Sheridan Gardiner single letters. Visuscopic fixation is R.E. parafoveal, nasal and superior and L.E. central to parafoveal superior.

Congenital esotropia, aged approx. 2 months.

Right micro-tropia, aged 3 years.

Results

Reponse to occlusion and increased abduction with the patient described has been most unusual and has avoided surgical correction so far. When occlusion was reduced to part-time and alternating, at the age of 2 years, the angle of deviation reduced dramatically, that is at that time the two eyes were able to function simultaneously for longer periods. Finally, the deviation reduced to a small esotropia of 11 prism dioptres, eliciting gross stereopsis (synoptophore). Abduction increased well at an early stage of management, and is now full. Bilateral overaction of the inferior oblique muscles, which was evident when abduction became full and easy, may present an obstacle to continued development of binocular function.

In my experience, results of early treatment have not been of this nature. This infant, where early occlusion was almost constant, did not seem to have suffered by deprivation of development of binocular pathways, as suggested by Hubel and Wiesel[4]. Also the evidence at present is that this child did not experience any form of binocular function prior to the onset of strabismus, which the parents are adamant was congenital. I consider that increased abduction certainly encouraged the reduction of the angle.

CONCLUSION

From the case discussed and other literature available, one realises that active therapy, at the earliest possible age is of tremendous advantage for the patient. One can postulate that once the suppression scotoma is reduced and there is a change in the motor adaptations as abduction becomes a full and easy task, conditions are favourable for encouragement of the desire for single binocular function. If the fusion faculty is present as an innate reflex at birth, the question arises as to why there is not a higher percentage of congenital esotropes gaining normal or abnormal binocular function when treated adequately at an early age?

There are two schools of thought re the prognosis of the congenital esotrope:

1. The earlier the straightening of the eyes, the more chance there is of functional development.
2. The congenital esotrope has almost no chance of gaining any functional cure, therefore early surgery is not always considered.

The described atypical response to early occlusion therapy and ocular mobility supports the former theory. That is, when the infant's eyes are in a straight or micro-tropic position, functional binocular vision may be established. That is, this innate fusion reflex may be developed some time after birth.

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