CONGENITAL PTOSIS WITH SYNCHRONOUS MOVEMENTS OF THE AFFECTED LID AND LOWER JAW.

BY

ALVIN A. HUBBELL, M.D.,

PROFESSOR OF OPHTHALMOLOGY IN THE MEDICAL DEPARTMENT OF NIAGARA UNIVERSITY, BUFFALO, N. Y.

(*With three illustrations.*)

Reprinted from the ARCHIVES OF OPHTHALMOLOGY, Vol. xxii., No. 1, 1893.

CONGENITAL PTOSIS WITH SYNCHRONOUS MOVEMENTS OF THE AFFECTED LID AND LOWER JAW.

By ALVIN A. HUBBELL, M.D.,

PROFESSOR OF OPHTHALMOLOGY IN THE MEDICAL DEPARTMENT OF NIAGARA UNIVERSITY, BUFFALO, N. Y.

(*With three illustrations.*)

T. W. B. is a boy now seven years of age. He is one of a family of three children whose parents are healthy and intelligent Americans. No malformation is present in any of them, except himself, and none is known to have existed in either the paternal or maternal ancestors. This boy is well developed, healthy, and unusually intelligent. He has a symmetrical and well-shaped head and face, and were it not for the ptosis on the right side he would be quite a handsome lad. Soon after birth it was noticed that the right eyelids did not open with the left, but would always remain closed, excepting when the mouth was opened, and then they would separate and close again with the closure of the mouth. This condition has continued without any apparent change to the present time. On examination I find that when the boy's mouth is closed the right eyelids are also closed, and there is not the slightest movement of the upper lid when the left eyelids are ordinarily opened, or the left eyeball is directed upwards, or when any other movement is made by which the eye or head is turned, or the face drawn, or the lower jaw carried to one side. Figure 1 shows the appearance of the boy's eyes when he is looking forward, with his mouth closed. If the boy is asked to open both eyes as widely as he can, the right upper lid is raised a little, as shown in Figure 2. But this is done entirely by the action of the occipito-frontalis muscle and not in the least by the

Reprinted from the ARCHIVES OF OPHTHALMOLOGY, Vol. xxii., No. 1, 1893.

levator palpebræ. When, however, the mouth is opened, the right upper lid is involuntarily and unconsciously raised at once. The greater the depression of the lower jaw the higher is the upper lid lifted. In other words, the contraction of the right levator muscle takes place involuntarily with the contraction of the depressor (digastricus) of the lower jaw, and exactly proportion-

FIGURE 1.

ately to that of the latter. This action, however, can be overcome by the orbicularis palpebrarum, whose function is normal on both sides, since the right eye can be voluntarily kept closed while the mouth is opened. When the right eyelids are separated, the eye is seen to be of the same size, and the iris of the same color as the left ; but there is marked divergent strabismus together with

some downward deviation. The eye cannot be turned upwards beyond the median plane, or inwards but little beyond the vertical plane; but downwards and outwards its movements are of usual, or perhaps more than usual extent. The pupil and iris are normal in appearance, size, and reflexes, and are like the left. The refraction of both eyes is, by the ophthalmoscope, emmetropic.

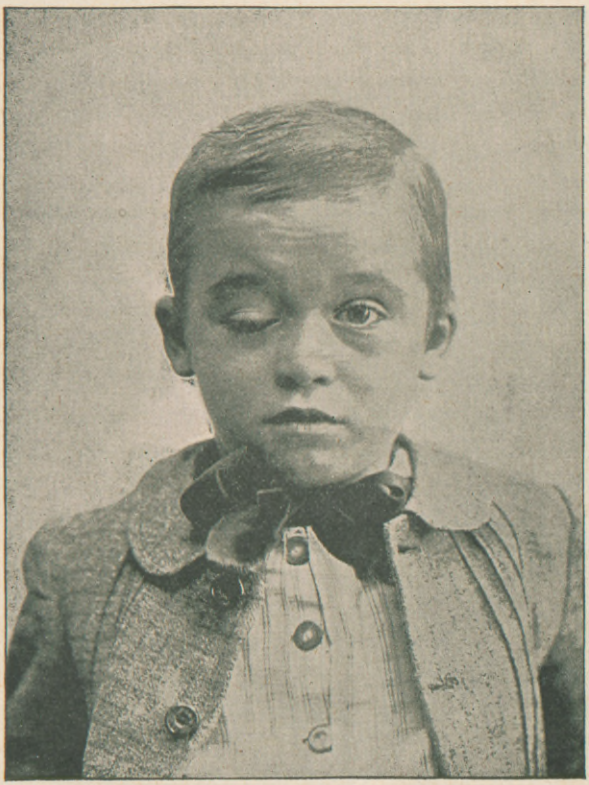

FIGURE 2.

The fundus of the right eye is, like the left, normal in appearance in every respect. But the vision of this eye is amblyopic, being able barely to count fingers at four feet, while that of the left is normal, reading number 6, Snellen, at six metres ($\frac{6}{6}$). Figure 3 shows the appearance of both eyes when the mouth is widely opened.

Remarks.—So far as I know, fifteen cases of this anomaly have been reported. While they have differed somewhat in some particulars, they have all had ptosis on one side with rising of the drooping lid synchronously with some movement of the lower jaw, generally depression. In no case has a post-mortem and histological examination of the brain

FIGURE 3.

been made. Hence the anatomical basis of these anomalous movements is at present merely conjectural. But taking into consideration the fact that the innervation for the levator palpebræ and the digastricus or other muscle of the lower jaw is in these cases undoubtedly one and the same, as shown by the associated movements of these muscles,

and that a part of the nucleus of the descending motor root of the fifth and the nucleus of the root of the third nerve are in close proximity to each other in the neighborhood of the aqueduct of Sylvius, the supposition becomes very probable that an abnormal connection is here established. This abnormal nuclear transposition may vary in different cases, but it appears always to be from some part of the third to some part of the descending root of the fifth nerve. A lack of proper development of the nucleus of origin of more or less fibres of the third nerve appears also to be a prominent characteristic in all cases. In my own case I believe that the origin of the fibres supplying the levator muscle has been transferred from the nucleus of the third nerve to that part of the nucleus of the fifth which sends fibres to the anterior belly of the digastric muscle. The nucleus of the third nerve seems to be undeveloped in that portion from which arise the fibres supplying the levator of the upper lid, the superior rectus, and the internal rectus muscles, while that part sending fibres to the inferior rectus (possibly the inferior oblique) and the iris is normally present. Thus do we get in this case the ptosis, the divergent strabismus, the downward deviation, and the associated movements of the upper lid with the lower jaw.

The Knickerbocker Press, New York
G. P. Putnam's Sons