POST-TRAUMATIC PULSATING EXOPHTHALMOS

CAUSED BY PERFORATION OF AN ERODED ORBITAL ROOF BY A HYDROCEPHALIC BRAIN

H. VERBIEST, M.D.*

University Hospital, Utrecht, Holland

(Received for publication October 8, 1952)

The relation between certain vascular anomalies and pulsating exophthalmos has been known for many years. The first fundamental observations were made by Travers (1809), Dalrymple (1815), Baron (1836) and Nelaton (1856). However, a long time elapsed before the importance of abnormal cranio-orbital communication as a cause of pulsating exophthalmos was clearly recognized. The publications of Moore, LeWald and Wheeler directed attention to pulsating exophthalmos in individuals with congenital defects of the superior or/and posterior orbital walls. In several of their cases the lesion was associated with diffuse neurofibromatosis.

Abnormal communication between the cranial cavity and the orbit may exist through preformed openings (foramina and fissures) and through congenital or acquired defects in the bony orbital walls. The acquired defects are traumatic or caused by disease. Strandberg divided the latter in order of frequency into: bone erosion caused by aneurysms, malignant tumours, inflammatory processes, angiomias, von Recklinghausen’s disease, Paget’s disease, and internal hydrocephalus. From the literature it appears that in such cases of pulsating exophthalmos the intracranial pressure may be normal.

Van der Hoeve published a case in which spontaneous pulsating exophthalmos was thought to be the consequence of internal hydrocephalus. According to him the increased intracranial pressure had caused a widening of the optic foramen, followed by a herniation of the brain through this opening.

It is most remarkable that operative removal of large parts of the orbital walls, which is inevitable in transcranial extirpation of intra-orbital tumours, is practically never complicated by pulsating exophthalmos. In his monograph on orbital tumours Dandy stated: “The thought doubtless occurs whether pulsating exophthalmos does not follow removal of the orbital roof. I have looked carefully for such a sequela but have never found it, despite the fact that pulsating exophthalmos is known to occur when the orbital roof is congenitally absent.”

Wheeler considered that the nature of the tissues filling the orbital defect determines whether the cerebral pressure and pulsations will be transmitted to the orbital content or not. After operative removal of the orbital

* Senior Neurosurgeon.
roof newly formed tissue replaces the defect and in the course of time this layer becomes firm connective tissue, acting as an effective barrier against intracranial pressure and pulsations.

In the writer’s experience, operative removal of the orbital roof was followed by pulsating exophthalmos in one instance. In this case the intracranial operative removal of the orbital roof was part of the treatment of a cranio-orbital injury. The postoperative course was complicated by meningitis and postmeningeal hydrocephalus. During the period of increased intracranial pressure pulsating exophthalmos developed, but it subsided completely after the successful relief of the hydrocephalus.

It therefore seems reasonable to distinguish the mechanisms of pulsating exophthalmos in cases of abnormal cranio-orbital communications in two categories: (1) A lowered mechanical resistance of the cranio-orbital barrier, which replaces the defect, either as a result of a developmental disturbance or local disease. It does not seem to be of fundamental importance that the intracranial pressure should be increased at the time of the examination. (2) The increased intracranial pressure and pulsations are the main factors against which the reparative processes replacing the orbital defect are only partly effective. There is a direct relation between the pulsating exophthalmos and increased intracranial pressure, viz. lowering of the intracranial pressure makes the exophthalmos disappear.

It is the second group that has aroused our special interest. In 2 of our cases the pulsating exophthalmos that occurred after cranial injuries resulted from herniation of a hydrocephalic brain through a perforation in the orbital roof.

In chronic internal hydrocephalus the thinning of the orbital roof from the pressure of the brain may be considerable. Fig. 79 in Dorothy Russell’s report on hydrocephalus shows marked erosions at the base of the skull in such a case; the bone of the orbital roof was so thin that a hole was accidentally made with the point of the forceps whilst stripping the dura, exposing underlying adipose tissue. If, for instance, such a person falls and strikes his head against something, the brain is consequently set into motion. If the movement of the skull is stopped, the brain continues its movement until this is resisted by certain parts of the skull. If the circumstances of the accident determine a rotatory movement of the brain towards the roof of the orbit, the latter yields to the impact, which means perforation of the roof and herniation of the brain into the orbit. It seems also possible that the fracture of the orbital roof is caused by vibrations, set up by the injurious forces. Sooner or later the fractured part of the orbital roof gives way to the high pressure of the intracranial content and intra-orbital herniation of the brain follows.

CASE REPORTS

Case 1. #N. 2080. Y.M., a female 24 years old, was admitted on July 22, 1946. In December 1943 she had been hit by a motorcar and was admitted to an Indonesian hospital with a basal fracture of the skull. She remained unconscious for 4
days. The right eye was displaced downwards and during subsequent years a progressive pulsating exophthalmos developed. Other disturbances such as amenorrhoea, bradypsychia and a certain stiffness of gait also remained.

Examination. There was bilateral papilloedema. The right eye protruded and was displaced downwards. Upward movement of this eye was limited. The cornea and sclera appeared normal; there was no chemosis. On palpation a soft mass above the eye was felt through the upper eyelid. No vascular murmur could be detected. There was dorsi-extension of the great toe on both sides (Babinski). Sugar toler-

ance test showed a normal fasting level, but the peak of the curve was 204 mg. per cent; a normal value, however, was restored within 2 hours. There was no abnormal reaction of the blood sugar to adrenalin and insulin. B.M.R. was −23 per cent. Water balance was normal.

Roentgenograms of the skull showed increased convolutional impressions. The cranial vault was very thin. The sella was enlarged with widening of the entrance. There was a depressed fracture of the right orbital roof (Fig. 1).

Ventriculography on Aug. 16, 1946, revealed a pronounced internal hydrocephalus and atresia of the aqueduct. The right frontal horn extended downwards into the right orbit (Fig. 2).

Operation. A right frontal craniotomy was performed. The dura mater was incised after ventricular tap. On elevating the tip of the frontal lobe, the prolapse into the orbital roof became visible. As this was irreducible, the neck was cut, whereby the ventricle was inevitably opened. The frontal lobe was then elevated, and using the procedure of Stookey and Scarff the lamina terminalis and the floor of
the 3rd ventricle were perforated to relieve the obstructive hydrocephalus. The orbital roof was exposed extradurally, the hernial formation was excised and the depressed bone was removed. The orbital roof was covered with an osteoperiosteal bone graft obtained from the tibia. The dural defect was closed with a fascia lata graft and the wound was closed.

**Course.** Postoperatively the eye did not protrude and there were no pulsations, although there was still some downward displacement of the eye. Convalescence was slow because of the patient’s attitude of indifference and inactivity, which had been apparent before operation.

![Image](image.jpg)

**Fig. 2. Case 1.** The frontal horn of the dilated right lateral ventricle shows an outpouching through the right orbital roof. The occluded aqueduct tapers to a point.

**Case 2.** #N. 8760. J.C. de K., a male 25 years old, had fallen from his bicycle on Aug. 31, 1951 and knocked his head against the tramway-rails. He visited his doctor and then went back to work. A few hours later, however, he began to feel ill and was admitted to hospital, where a diagnosis of basal fracture of the skull and a pulsating exophthalmos of the right eye was made. On Sept. 2, 1951, he was transferred to the neurosurgical ward as it was suspected that an arteriovenous fistula was the cause of the pulsating exophthalmos.

**Examination.** The patient complained of attacks of headache and vomiting. There was an extreme pulsating exophthalmos of the right eye, with chemosis and keratitis. The eye was deviated downward. All eye movements were limited, especially elevation. A soft mass was found protruding behind the upper eyelid. This mass could be reduced by finger pressure and it did not seem to be adherent to the eyebulb.

Roentgenograms of the skull revealed enlargement and erosion of the sella. The occipitomental view showed a fracture with perforation of the anterior part of
the right orbital roof (Fig. 3); the occipitofrontal view did not show these orbital changes.

Ventriculography was done with a small quantity of air according to the method of Ziedses des Plantes. Both lateral ventricles were dilated; after the "saltus" the air collected in a widened posterior part of the 3rd ventricle; the anterior part of the aqueduct was filled and it was not funnel-shaped. The exophthalmos could be reduced by lowering the ventricular pressure.

1st Operation, Sept. 3, 1951. Our first attempt was to relieve the hydrocephalus. As there was some doubt about the nature of the obstruction the approach was made by a suboccipital craniotomy. The arch of the atlas was removed in order to free the incarcerated tonsils of the cerebellum. The cerebellum appeared completely normal and the 4th ventricle was intact. A No. 10 catheter was left in the aqueduct for some days with the other end of it guided through the muscles and skin, and connected with a receptacle. The wound was closed.

Course. After the operation there was only a slight degree of exophthalmos with faint pulsations.

On Sept. 6, 1951 there was no outflow from the catheter. The intracranial pressure was high (35 cm. H₂O) and there was pronounced exophthalmos. The catheter was withdrawn and the ventricle was tapped. The patient became soporose. Arterial hypertension, bronchial hypersecretion and profuse sweating developed.

2nd Operation. The aqueduct and the 4th ventricle were found to be blocked by blood clots, which were removed. Lavage of the ventricles was performed. whereupon the CSF circulation seemed to be re-established.
Course. The ventricular pressure remained normal, and the exophthalmos completely disappeared. However, the keratitis, present on admission, had resulted in panophthalma, necessitating evisceration of the bulbus on Sept. 14, 1951. Thereafter convalescence was uneventful.

3rd Operation. Two months later, on Nov. 12, 1951 right frontal craniectomy was done. The cerebral prolapse into the orbit appeared to be irreducible. The prolapse was divided at the neck and the intra-orbital part was excised. The orbital roof was covered by an osseous graft from the ilium and the dural defect was closed by a fascia lata graft.

Course. The patient left hospital 2 weeks later with a normal intracranial pressure. No pulsations could be felt through the orbit.

DISCUSSION

Although nowadays it is generally known that pulsating exophthalmos may be produced by different lesions, many clinicians are still inclined to attribute all cases of post-traumatic pulsating exophthalmos to a carotid-cavernous aneurysm (Rowbotham; Mock). In discussing the differential diagnosis, Mock stated: "A history of trauma will nearly always be obtainable for the diagnosis of a carotid-cavernous arteriovenous aneurysm." Our 2 cases show that traumatic pulsating exophthalmos may be caused by another condition, namely perforation of a previously eroded and thin orbital roof by the impact of a hydrocephalic brain. These 2 cases represent 28 per cent of the total number of cases of post-traumatic pulsating exophthalmos observed in our hospital during the last 10 years. In our other 5 cases the cause was an arteriovenous aneurysm.

DIAGNOSIS

In order to avoid a diagnostic error in cases of post-traumatic pulsating exophthalmos, it is very important to make an X-ray examination of the skull, especially of the orbital roof. Sometimes the defect may not be visible in the occipitofrontal view of the skull, whereas it is usually evident in the occipitomental view (see Case 2). Post-traumatic pulsating exophthalmos caused by hydrocephalus and perforation of the orbital roof may have the following features in common with the exophthalmos associated with arteriovenous aneurysm:

1. The pulsations of the eye disappear or diminish on compression of the carotid artery.
2. Chemosis of the affected eye (Case 2) may be present.
3. Papilloedema. It should be noted, however, that in Case 1 papilloedema was present not only on the affected side, but was bilateral.

In post-traumatic pulsating exophthalmos caused by herniation of hydrocephalic brain there are no vascular thrills or murmurs. The protruding brain may be visible or palpated behind the upper eyelid. The exophthalmos diminishes after a tap of ventricular fluid. The skull may show typical signs of chronic hydrocephalus. Ventriculography serves as a most valuable aid in the diagnosis.
It should also be pointed out that the patient may never have noticed a pre-existing pulsating exophthalmos associated with a congenital orbital defect and that his attention was drawn to it only after the trauma. This may give a wrong impression to the doctor who examines the patient for the first time after the trauma.

Treatment. The first objective should be to relieve the internal hydrocephalus, either by removal of the cause of obstruction or by palliative procedures, depending upon the circumstances. Once the intracranial pressure has been reduced, the treatment of the cerebral hernia with plastic repair of the orbital defect will not be found difficult.

Failure to recognize this condition may lead to erroneous interventions, such as carotid ligation. Gardner reported a case of post-traumatic unilateral exophthalmos caused by outpouching of a hydrocephalic brain through a fractured orbital roof. The patient suffered from a cerebellar tumour. A frontal craniotomy was performed, as a diagnosis of meningioma of the sphenoid ridge with invasion of the orbit had been made. The operation revealed the true nature of the lesion, and further examination led to the surgical treatment of a large cerebellar cystic haemangioblastoma, which had caused the obstructive hydrocephalus.

Corbella described a 26-year-old patient with post-traumatic exophthalmos. Enucleation of the eye was followed by an enormous intra-orbital protrusion, which became visible in the region of the upper eyelid. This protrusion consisted of herniated frontal lobe containing a diverticulum of the ventricle. Roentgenograms of the skull showed convolutional markings and separation of the sutures. Ventriculography demonstrated an internal hydrocephalus. An effort to reduce the cerebral herniation by a transfrontal approach failed and the patient died 2 days later. Corbella thought that the traumatic brain lesion was the cause of the hydrocephalus. However, it seems more probable that the changes in the skull evident in the X-rays were not the result of hydrocephalus of 3 months' duration, as reported by the author, but were caused by chronic pre-existing hydrocephalus. From this point of view Corbella's case might be considered in the category of patients with chronic hydrocephalus, in whom a cranial injury results in traumatic rupture of the orbital roof with outpouching of the brain into the orbit.

SUMMARY

Although abnormal cranio-orbital communications are well-known causes of pulsating exophthalmos, there is still a general tendency to attribute all cases of post-traumatic pulsating exophthalmos to vascular lesions. The author, however, points to a special type of post-traumatic pulsating exophthalmos that occurs in patients in whom the orbital roof has become thin and eroded as a result of chronic hydrocephalus. The secondary movements of the brain and of the ventricular fluid, set up by the injury, may force the brain through the weakened orbital roof, or the vibrations, set up by the injurious forces, may result in a fracture of the orbital roof, present-
ing a locus minoris resistentiae to the expansive forces of the hydrocephalic brain. If the patient had had no complaint indicative of chronic hydrocephalus prior to the injury, the danger of a diagnostic error is imminent. Every patient suffering from post-traumatic pulsating exophthalmos should be subjected to a thorough X-ray examination of the orbital roof in different views. Other valuable diagnostic aids are the roentgenological signs of chronic hydrocephalus (ventriculography included) and the relation of ventricular pressure to the degree of exophthalmos.

REFERENCES
4. HOEVE, J. VAN DER Cited by Strandberg.15