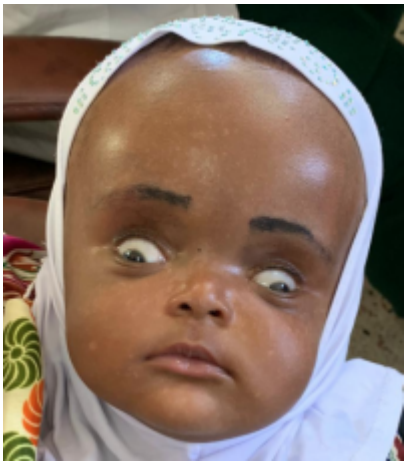
Setting sun sign

J.Sales-Llopis

Neurosurgery Service, Alicante University General Hospital, Alicante Institute for Health and Biomedical Research (ISABIAL - FISABIO Foundation), Alicante, Spain.



The setting sun sign (also known as the sunset eye sign or setting sun phenomenon) is a clinical phenomenon encountered in infants and young children with raised intracranial pressure.

It is an earlier sign of hydrocephalus than enlarged head circumference, full fontanelle, separation of sutures, irritability, vomiting. Consequently, this sign is a valuable early warning of an entity requiring prompt neuroimaging and urgent surgical intervention [1)].

Part of Parinaud's syndrome.

Definition

The "setting sun" sign is an ophthalmologic phenomenon where the eyes appear driven downward bilaterally. The inferior border of the pupil is often covered by the lower eyelid, creating the "sunset" appearance. This finding is classically associated with hydrocephalus in infants and children.

Epidemiology

Seen in up to 40% of children with obstructive hydrocephalus and 13% of children with shunt dysfunction [2)].

In 126 children with internal hydrocephalus setting sun was descibed in 51, syndrome of the aqueduct of Sylvius 14, paresis of craniocerebral nerves 9, nystagmus 8, optic atrophy 4 [3)].

Clinical features

It consists of an up-gaze paresis with the eyes appearing driven downward. The lower portion of the pupil may be covered by the lower eyelid, and sclera may be seen between the upper eyelid and the iris.

Pathogenesis

The pathogenesis of the setting sun sign is believed to be related to aqueductal distention in the dorsal midbrain on the vertical gaze innervation bilaterally. In children with hydrocephalus, up to 40% of cases will present with this sign. Of these patients, 13% harbor ventriculoperitoneal shunts that have failed. The sign is also associated with kernicterus and other features of the full Parinaud syndrome (i.e., dorsal midbrain syndrome). Interestingly, the setting sun sign may also transiently appear in healthy infants up to 7 months of age [4)].

Chattha et al. suggest periaqueductal dysfunction rather than mechanical displacement as the possible mechanism for this sign [5)].

Outcome

In hydrocephalus, the convulsion and so-called setting sun sign had no significant correlation to poor prognosis [6)].

Despite the fact that setting sun eye is a grave sign, most commonly accompanied by other neurological signs and symptoms suggesting serious diseases, it might be observed as a sole finding in a totally normal infant with inconclusive brain imaging and laboratory tests [7)].

Case series

A cross-sectional study was conducted in the Children's Hospital Medical Center in Tehran from June 2001 to 2006. The study included 15 healthy infants who were referred to the neurosurgery clinic for setting sun eye. All were evaluated with brain imaging, and laboratory tests including at least thyroid function tests, and serum calcium and phosphorus. The cases were followed by regular outpatient visits until the age of 2 years.

They were 3-8 months old at the time of referring to the outpatient clinic. Setting sun eye was observed by the mother in all cases and confirmed during their visit to the clinic. All had normal brain imaging and normal laboratory tests (thyroid function and electrolytes). Setting sun eye disappeared gradually during the follow-up period with a range of 2-8 months after detection by the mother.

Despite the fact that setting sun eye is a grave sign, most commonly accompanied by other neurological signs and symptoms suggesting serious diseases, it might be observed as a sole finding in a totally normal infant with inconclusive brain imaging and laboratory tests. We found that this type of setting sun eye has a benign course and will disappear without any intervention several months after its detection (commonly before the age of 2 years without any intervention) [8)].

19 infants who displayed the setting-sun eye phenomenon were observed during the first year of life. Nine of the infants showed no signs of illness, eight had an evident increase in intracranial pressure requiring surgical relief, and two had transient signs of increased intracranial pressure which resolved spontaneously. The setting-sun phenomenon could be elicited both by alteration of the infant's position and by removal of light, and it also occurred spontaneously. The effectiveness of the eliciting mechanism depended on the age of the infant. The component parts of the phenomenon consist of downward rotation of the eyeballs and retraction of the upper eyelids, sometimes accompanied by raising of the brow. The phenomenon can be observed in healthy infants, and its value in early recognition of increased intracranial pressure is limited. The response might indicate increased intracranial pressure if it can be elicited by alteration of position in infants older than four weeks of age or if there is a marked response to removal of light in infants younger than eight weeks or older than 20 weeks of age, especially if the response is combined with constant or intermittent strabismus or undulating eye-movements [9].

Eight cases of obstructive hydrocephalus manifesting palsy of upward gaze and other features of the Sylvian aqueduct syndrome are reported. During the crisis of intracranial hypertension, all of them developed upward gaze palsy and variable abnormalities of the convergence mechanism such as paralysis, spasm, and convergence nystagmus. The frequent apparent blindness was probably related to gaze paralysis since visual evoked responses were present. All these ocular abnormalities disappeared after shunting. Periaqueductal dysfunction on the basis of raised intracranial pressure is postulated as the possible mechanism for the above ocular manifestations. The 'setting sun' sign is frequently seen in infants and children with hydrocephalus and has been considered in the past to result from the displacement of eyeballs by pressure from the orbital roof plate. Our observations would suggest periaqueductal dysfunction rather than the mechanical displacement as the possible mechanism for this sign [10].

Case reports

Yoshikawa reported two normally developed infants showing benign“ setting sun” phenomenon. A 2(2-12)-year-old boy and a 7-year-old boy, who were born without any complications at full term, developed brief episodes of downward gazing during sucking and crying after birth However, there were no other clinical or laboratory findings, and they developed normally. The phenomenon was not visible until 6 months and 7 months, respectively. The “setting sun” phenomenon usually indicates underlying severe brain damage and can also be seen, although rarely, in healthy full-term infants until 1 to 5 months. However, the benign “setting sun” phenomenon might exist until 6 or 7 months of age in normal infants [11].

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