EDITORIAL

The "Setting Sun" Sign: A proposed Grading Scale

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ABSTRACT:
The "Setting Sun" sign is a neuro-ophthalmic finding observed in infants with hydrocephalus, characterized by conjugate downgaze due to pressure effects on the posterior third ventricle and midbrain. This sign can be perceived in about third of infants with obstructive hydrocephalus, and could serve as an early clinical indicator for the progression in intracranial pressure. Despite its significance, there is a paucity of standardization to quantify the hydrocephalus severity, limiting objective assessment and monitoring. Here, we propose a novel, simple and sensitive grading system based on the degree of iris and pupil coverage by the lower eyelid. This system aims to enhance early detection, facilitate disease monitoring, and improve treatment decision-making in the management of infantile hydrocephalus.

KEYWORDS: Setting-Sun sign, infantile obstructive hydrocephalus.

Abbreviations: MRI: Magnetic Resonance Imaging.

INTRODUCTION:

Clinical measurement scales play a crucial role in evaluating patient status and tracking disease progression. Standardized tools such as the Glasgow Coma Scale[1], Karnofsky Scale[2], Rankin Scale[3] and Thyroid Exophthalmos Scale enable quantitative assessments, improving clinical decision-making. However, no formal grading system exists for the "Setting Sun" sign, a key neuro-ophthalmic marker of hydrocephalus first described by Cernerud in 1975[4]. This phenomenon, characterized by downward eye deviation and superior scleral exposure, occurs due to pressure effects on the midbrain, leading to impaired upward gaze. It is commonly associated with hydrocephalus but can also appear in conditions such as Parinaud's syndrome[5], kernicterus[6], and transiently in some healthy infants[7]. It is encountered in infants and young children with raised intracranial pressure (seen in up to 40% of children with obstructive hydrocephalus and 13% of children with shunt dysfunction)[8].Given the dynamic nature of the "Setting Sun" sign, a standardized grading system is necessary to improve early diagnosis, track progression, and guide timely interventions.

In the normal infant, the upper lid covers the upper part of iris, while in hydrocephalus, the lower eyelid covers the iris at various degrees depending on the severity. Given the dynamic nature of the "Setting Sun" sign, where severity fluctuates with hydrocephalus progression or improvement following interventions such as a ventriculoperitoneal shunt, an objective grading system is necessary. Such a system would:

1. Improve Early Detection – Allow clinicians to document subtle changes in ocular findings before hydrocephalus worsens.
2. Facilitate Monitoring – Enable consistent tracking of disease progression and response to treatment.
3. Enhance Decision-Making – Assist neurosurgeons and pediatricians in determining when surgical intervention (e.g., shunt placement or revision) is required.
4. Promote Research and Clinical Communication – Establish a universal reference point that can be used in studies and clinical reports to describe the severity of the "Setting Sun" sign more precisely.

The proposed "Setting Sun" sign grading system:

In a normal anatomical state, the upper eyelid typically covers the upper part of the iris, leaving only a portion of the sclera visible inferiorly. This position is maintained by the coordinated function of the levator palpebrae superioris muscle and Müller's muscle, which regulate upper eyelid positioning. Additionally, normal eye movements allow for smooth conjugate gaze in all directions, including unrestricted upward gaze. In contrast, the 'Setting Sun' sign is characterized by downward deviation of the eyes, exposing an abnormally large portion of the superior sclera. This occurs due to upper lid retraction and impaired upward gaze, a hallmark of midbrain dysfunction commonly seen in hydrocephalus. The lower eyelid progressively covers more of the pupil and iris as the severity of the condition increases. Understanding the normal eye position is essential in recognizing and grading this pathological deviation.

To standardize the assessment of the "Setting Sun" sign, we introduce a five-grade classification system based on the degree of iris and pupil coverage by the lower eyelid. This scale can serve as a quantitative tool for monitoring patients, particularly in cases of shunt failure or progressive hydrocephalus.

Grade 0 (Normal Eye Position): The upper eyelid covers the upper portion of the iris, and the eye maintains normal upward gaze ability.

Grade I The lower eyelid covers only the lower part of the iris (mild increase in intracranial pressure)..

Grade II The lower eyelid reaches the lower edge of the pupil.

Grade III The lower eyelid covers the lower half of the pupil.

Grade IVOnly the upper part of the pupil is visible. (Moderate-to-severe hydrocephalus).

Grade VOnly the upper tip of the iris is visible (severe hydrocephalus requiring urgent intervention).

Where grade I is the early increase in the intracranial pressure and the higher the grade may indicate worsening hydrocephalus.

Table 1: The "Setting Sun" sign grading system with illustrative depiction and real case examples.

"Setting Sun" Sign Grading Scale			
Grade	**Description**	**Depiction**	**Real Case Samples**
Grade 0	**(Normal) Upper lid covers upper iris**		
Grade I	**Lower lid covers lower part of iris**		
Grade II	**Lower lid touches lower edge of pupil**		
Grade III	**Lower lid covers half of pupil**		
Grade IV	**Only upper part of pupil is showing**		
Grade V	**Only upper tip of iris is showing**		

This grading system provides a structured approach to evaluating the severity of the "Setting Sun" sign, making it a potential tool for identifying early signs of hydrocephalus decompensation. Future studies should assess its inter-rater reliability and compare its findings with neuroimaging results, such as This grading system provides a structured approach to evaluating the severity of the 'Setting Sun' sign, making it a potential tool for identifying early signs of hydrocephalus decompensation. Future studies should assess its inter-rater reliability and compare its findings with neuroimaging results, such as ventricular index measurements or optic nerve sheath diameter changes on MRI. By integrating this scale into routine clinical evaluations, clinicians may achieve earlier detection of hydrocephalus worsening, improving patient outcomes.

This grading system allows for a more structured approach to evaluating the severity of the "Setting Sun" sign, making it a potential tool for identifying early signs of hydrocephalus decompensation. Future studies should assess its inter-rater reliability and compare its findings with neuroimaging results, such as ventricular index measurements or optic nerve sheath diameter changes on MRI.

Limitations:

This grading scale is based on clinical observations that may vary between patients due to individual differences in eyelid position and ocular muscle tone. It is not intended to replace radiological assessment but rather to complement existing diagnostic tools. Further prospective studies are needed to evaluate its inter-rater reliability, correlation with imaging findings, and potential role in predicting hydrocephalus progression or shunt failure.

CONCLUSION:

With over three decades of experience managing pediatric hydrocephalus, we propose a novel grading system for the "Setting Sun" sign. This standardized classification enables more precise monitoring of hydrocephalus progression and treatment response. Future research may address the accuracy, and the reliability of this grading system, and could investigate the potential correlation with neuroimaging and clinical assessment. Once validated, this system could enhance early diagnosis, improve clinical communication, and optimize treatment strategies for infants with hydrocephalus.

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REFERENCES:

1. Teasdale G, Jennett B. Assessment of coma and impaired consciousness: a practical scale. The Lancet. 1974;304(7872):81-4.
2. Karnofsky DA. The clinical evaluation of chemotherapeutic agents in cancer. Evaluation of chemotherapeutic agents. 1949:191-205.
3. Broderick JP, Adeoye O, Elm J. Evolution of the modified Rankin scale and its use in future stroke trials. Stroke. 2017 ;48(7):2007-12.
4. Cernerud L. The Setting-sun Eye Phenomenon in Infancy. Developmental Medicine & Child Neurology. 1975 ;17(4):447-55.
5. Ortiz JF, Eissa-Garces A, Ruxmohan S, Cuenca V, Kaur M, Fabara SP, Khurana M, Parwani J, Paez M, Anwar F. Understanding Parinaud's Syndrome. Brain Sci. 2021, 11, 1469 [Internet]. 2021
6. Das S, van Landeghem FK. Clinicopathological spectrum of bilirubin encephalopathy/kernicterus. Diagnostics. 2019 ;9(1):24.
7. Nejat F, Yazdani S, El Khashab M. Setting sun eye in normal healthy infants. Pediatric Neurosurgery. 2008 ;44(3):190-2.
8. Boragina M, Cohen E. An infant with the "setting-sun" eye phenomenon. CMAJ. 2006 ;175(8):878-.