exactitude, it behooves us to use a more precise term such as TAO, for the most commonly occurring orbital disease. In doing so, we seek not to remove diversity in the medical literature but to reduce sources of confusion for our colleagues and patients and in electronic searches.

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“Percussive” orbital trauma from wooden drumstick

Although penetrating orbital trauma are rare,¹ they are associated with high rates of mortality and monocular blindness.² In addition, they can cause damage to adjacent structures such as the brain, the cavernous sinus, and major arteries. Reported complications include hemorrhage, thrombosis, neurological deficits, facial fractures and associated deformations, and infections.¹ Optimal management of orbital penetrating injuries includes prompt and thorough evaluation and multidisciplinary collaboration between ophthalmology, neurosurgery, trauma surgery, otolaryngology, and radiology.

Here, we present a case of a 68-year-old man who sustained an orbital penetration injury with a 43 cm musical drumstick, which entered the right orbit, displaced the globe, and transited to the left maxillary sinus. Interestingly, the injury spared the globe, optic nerve, brain, and large blood vessels. Swift removal under endoscopic visualization was critical in the successful surgical management of the patient, and the patient’s vision was unaffected. To the best of our knowledge, no reported case of penetrating orbital trauma has involved a foreign body of this length without the occurrence of permanent visual or neurologic sequelae.

A 68-year-old man presented as a trauma transfer with a penetrating orbital injury. According to emergency medical services, a wooden musical drumstick was used as a weapon to impale the right orbit of the patient by another individual during an altercation. The patient arrived at our institution intubated and sedated from an outside hospital, where tetanus prophylaxis had been administered. He was hemodynamically stable. Physical examination revealed a large wooden drumstick still present in the right orbit; the drumstick penetrated the medial aspect of the right lower eyelid, with its vector directed inferomedially toward the nose and the lower left side of the face. The right eye could not be visualized on attempt to open the eyelid. The left eye appeared normal and was reactive to light and accommodation. Computed tomography (CT) of the head revealed that the object pierced the right inferior and medial orbital walls at the level of the naso-orbito-ethmoid complex, and crossed obliquely through both ethmoid sinuses and the nasal cavity with the tip ultimately resting in the left maxillary sinus (Fig. 1). The estimated length of the intraorbital portion of the drumstick was 3.8 cm. The drumstick’s trajectory abutted the left sphenoid sinus, adjacent to but not penetrating the carotid (Supplementary Fig. 1, available online). No hematoma was noted. The trauma additionally produced an orbital floor fracture on the contralateral side.

Fig. 1—(A) External photograph depicting the position and trajectory of the musical drumstick in the right orbit. (B) Computed tomography image demonstrating the radiolucent wooden drumstick (blue asterisk) penetrating the medial orbital wall, and into the nasal cavities.
Fortuitously, the drumstick’s trajectory spared the right globe and optic nerve, with both appearing intact and displaced superiorly. Given the oblique trajectory of the weapon from left to right, the assailant was presumed to be left-handed, though this was unconfirmed.

Surgical extirpation of the drumstick was promptly performed the same day under direct visualization as well as with endoscopic visualization of the sinus anatomy via an endoscopic sinonasal approach. Endoscopic visualization of the object confirmed that it appeared intact in the sinuses without any structural irregularities or sharp edges. No sinonasal hemorrhage occurred during removal. Upon examination, the drumstick measured 43 cm in length (Supplementary Fig. 2A, available online). Inspection of the drumstick after removal revealed black electrical tape wrapped tightly near the tip—a protective measure typically employed by drummers to prevent use-related damage and increase the longevity of their instruments. The drumstick appeared completely intact upon removal, though there was some fragmentation of the black tape. Fragments of tape were noted anteriorly at the eyelid entrance wound and were removed with forceps. Endoscopic visualization confirmed no residual foreign bodies in the sinuses or nasal cavity. After removal of the object, orbital exploration was performed, where fragments of ethmoid bone were removed with forceps and no further drumstick elements were visualized. A solution of dilute bacitracin in saline was used to irrigate the orbit and then removed with suction.

Further exploration revealed no damage to the medial canthal tendon along the medial orbital wall, and probing demonstrated an intact lacrimal system. Forced ductions were negative, and globe exploration confirmed no evidence of rupture. The soft tissue lacerations were copiously irrigated and the lower eyelid was repaired. Prophylactic oral amoxicillin-clavulanate was prescribed to cover for nasal flora. The patient remained hemodynamically stable, and there was no bradycardia either at presentation or throughout the surgical proceedings. After 24 hours, the patient was clinically stable; vision was found to be at baseline and extraocular motility was full. Postoperative examination confirmed good apposition of eyelid tissues at the former penetration site (Supplementary Fig. 2B, available online) and a well-formed globe (Supplementary Fig. 2C, available online).

A multitude of stick-like foreign bodies have been reported to cause penetrating orbital injury—often with significant morbidity—including writing tools, weapons, metallic objects, and even animal appendages. These injuries can result in profound morbidity, including permanent loss of vision, and even death. Owing to the variety of possible presentations and complications, a comprehensive evaluation should be conducted. Radiographic and CT imaging of the head and neck should be performed to assess the object’s trajectory and evaluate the extent of anatomical damage, which may involve not just the adjacent nerves, blood vessels, and intracranial structures, but also the cervical vertebrae. Timely surgical exploration and removal of the foreign body by an interdisciplinary team should follow. It is important to consider that organic intraorbital foreign bodies can induce significant inflammation or infection, and therefore close postoperative monitoring is warranted for any retained fragments of the penetrating object.

The case presented herein is of a potentially devastating assault to the orbit with a distinct and strikingly long percussion tool. This “percussive” orbital trauma remarkably only injured an eyelid, the bony orbit, nasal cavity, and sinuses, without damaging any critical ocular, periorbital, orbital, sinus, or vascular structures. CT imaging was essential to rule out hematoma and vascular lesions, which may precipitate hemorrhage during removal of the foreign body, a commonly reported complication. No functional impairment or complications were observed in the postoperative period.

In addition to the uniqueness of the penetrating foreign body involved and the lack of subsequent visual impairment, this case is also notable as there has been no previously published report describing removal of a foreign body with assistance of endoscopic visualization. Although the trajectory of the drumstick was straight and predictable, endoscopy was used for visualization, as it was unknown if there were any small irregularities in its structure not visible on CT that could potentially injure vessels or other critical structures during extirpation. Further, endoscopy was critical to ensure that the distal drumstick was intact throughout the removal process, and to help rapidly identify any potential sinonasal hemorrhage occurring during or after removal. We recommend that this measure provides an additional dimension of visualization that can facilitate safe, complete removal, and increase the likelihood of favourable results.

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Supplementary Materials

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This report adhered to the ethics principles outlined in the Declaration of Helsinki as amended in 2013. The patient described herein provided written consent to publish identifiable photographs related to this case.

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Early ocular findings in Cohen syndrome: case report and Canadian survey study

Cohen syndrome (CS) is an extremely rare genetic disorder characterized by systemic and ocular findings, with fewer than 1000 cases estimated worldwide.1 The condition is caused by biallelic mutations in the vacuolar protein sorting 13 homolog B (VPS13B) gene.1 A diagnosis may be suspected in a child presenting with global developmental delay, hypotonia, microcephaly, slender hands and feet, neutropenia, and dysmorphic facial features (down-slanting palpebral fissures, hypertelorism, short philtrum, prominent upper teeth, maxillary hypoplasia, and micrognathia).2 Other typical facial characteristics in CS include thick eyebrows, thick bushy hair, low hairline, and long and thick eyelashes.3 Patients with CS have been described to be sociable with a cheerful disposition.1,2 Myopia and retinochoroidal dystrophy are very common, whereas strabismus, ptosis, and lens opacities are occasionally present.2 Owing to the rarity, nonspecific presentation, and apparent lack of family history, diagnosis may be delayed until later childhood.3 As such, reports of early findings in CS are scarce.4 Here, we describe 6-year longitudinal data from a child with CS beginning at day 1 of life. Motivated by this case, we additionally undertook a short survey study to determine the experience of pediatric ophthalmologists in Canada with this disorder.

A 1-day-old male born by C-section after uncomplicated term pregnancy was referred for ptosis. There was no contributory family history and no consanguinity. Systemic examination was normal. Ophthalmologic examination revealed isolated congenital left-sided ptosis. Anterior and posterior segments were normal. Ptosis was repaired at 2 months.

At 9 months, ptosis repair was stable but the retina demonstrated subretinal dystrophy. Refractive error was $+0.50 \times 90^\circ/ +0.50 \times 80^\circ$ (OD/OS). Assessment by general pediatrics at 10 months noted global delay, hypotonia, small mouth, and neutropenia. Magnetic resonance imaging of the head showed no evidence of microcephaly, and screening microarray was normal. At 29 months, cycloplegic refraction was $-4.75+0.50 \times 100^\circ/-4.50+0.50 \times 90^\circ$. Macular hyperpigmentation remained stable. Glasses were prescribed and resulted in improved visual behaviour. Uncorrected visual acuity improved from 20/190 OU to best-corrected visual acuity (BCVA) of 20/94 OU by Teller acuity cards.

Exome sequencing at 3 years identified 2 pathogenic variants of the VPS13B gene (c.1915C>T, c.6732+1G>A). On testing, both parents harboured one variant each, confirming the disease segregation that established the clinical diagnosis of CS. Refractive error was $-8.00+3.25 \times 115^\circ/-7.50+3.00 \times 80^\circ$ and BCVA was 20/63 OU. Fundus photographs demonstrated bulb’s eye macular hyperpigmentation with surrounding retinal elevation (Fig. 1a). Numerous rounded chorioretinal atrophic lesions with largely nummular pigmentary changes were present in the periphery (Fig. 1b). Macular optical coherence tomography (OCT) scans demonstrated bilateral large schiotic/cystoid changes in outer and inner nuclear layers with disruption of photoreceptor outer and inner segments in the parafoveal region in either eye and in the temporal macula of the left eye (Fig. 1c). Topical dorzolamide was initiated; however, it was abandoned owing to poor tolerance. Electroretinogram demonstrated severe rod-cone dystrophy (Fig. 2).

Subsequent assessments revealed myopic progression to $-10.50+6.50 \times 115^\circ/-10.00+5.00 \times 70^\circ$ at age 6 years. At this examination under anaesthesia, fundus photographs were stable (Fig. 1d, e). Fluorescein angiography showed no leakage at the macula, whereas the peripheral atrophic scars stained prominently (Fig. 1f). BCVA was 20/50 OU.

To our knowledge, this case is the earliest at which serial ophthalmologic assessments in CS have been reported. The earliest ocular findings of CS in this patient were congenital ptosis and macular hyperpigmentation first noted at 9 months, as well as early onset of progressive myopia requiring spectacle correction at age 2 years. Structural retinal changes on OCT, nonleaking cystoid macular edema (CME), and rod-cone dystrophy were detected after formal diagnosis at age 3 years, although may have been present earlier.

A previous case report of a 13-month-old child with CS described low myopia, abnormal foveal pigmentation, retinal cysts, and rod-cone dystrophy.4 Fluorescein angiography findings were not reported in that case, although nonleaking CME has been described in an 11-year-old with CS.3

Biallelic mutations in VPS13B identified in the proband confirmed the molecular basis of CS phenotype. The VPS13B gene mapped to 8q22.2 codes for multiple transcripts, the longest of which (NM_017890.4) is widely expressed in human tissues and thought to play a role in intracellular transport of proteins within Golgi complex. Knocking down VPS13B expression in the retinal pigment epithelium leads to abnormal glycosylation and might explain the retinal dystrophy seen in CS; however, further studies are needed to confirm this hypothesis.