Case Report of a Severe Recurrent Tongue Self-Injury in an Infant With Dystonia

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Dystonia is characterized by sustained or intermittent muscle contractions causing abnormal, often repetitive movements, postures, or both that are typically patterned, twisting, and sometimes tremulous. It is often initiated or worsened by voluntary action and associated with overflow muscle activation. In this article we report a case of severe oromandibular dystonia, which is a specific form of dystonia characterized by involuntary, action-induced tonic or clonic spasms of the masticatory, lingual, and pharyngeal musculature. Episodes of repeated tongue biting in a 17-month-old girl caused her to stay in the PICU for 4 weeks. These episodes were the consequence of dystonia induced by a perinatal stroke. We highlight the specific dental management that enabled us to treat the child without extractions. Facing this type of complex illness, we insist on the importance of interdisciplinary work with the goal of avoiding outdated techniques. The use of botulinum toxin seemed relevant.

CASE REPORT

C.V., a 17-month-old girl, was admitted to the pediatric emergency department of the University Hospital Center (CHU) of Bordeaux on April 14, 2015 for paroxysmal episodes of tongue biting ongoing for 4 days. Her medical history was marked by prenatal diagnosis of a right porencephalic cavity presumed to be related to stroke, right eye surgery (April 2014) linked to ptosis associated with a palsy of the third right cranial nerve, and left hemiparesis predominant in the upper limb. An EEG was performed on January 29 because of concerns that jerky head movements may have been due to epileptic seizures. These head movements had no ictal correlate and were thought to represent stereotypies.

On April 10 C.V. was admitted to a regional hospital because of an unusual bruxism and emesis quickly associated with the first episode of trismus with tongue biting. It was triggered by stimulation, inducing chewing and severe bleeding. On April 12, repeated episodes of tongue biting happened, preceded by upper limb hypertonia, clenched fists, lower limb extension (choreodystonic movements), and shouts. Antalgic treatment (acetaminophen and morphine) was not sufficient to terminate the episode. On April 13 hyperthermia at 39°C occurred; acyclovir was prescribed because of suspicion of encephalitis. Amoxicillin and clavulanic acid were added. The same day, an EEG was performed and showed large intermittent δ waves in right occipital region during sleep. Immediate-release diazepam (0.5 mg/kg) and then fosphenytoin sodium (15 mg/kg) were administered but failed. She was admitted on April 14 to

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Oral informed consent was obtained from the patient's legal guardians for publication of this case report and any accompanying images.

Drs Brissaud and Thébaud managed the clinical case and reviewed and revised the manuscript; Drs Guichoux and Villega managed the clinical case; Rawen Smirani drafted the initial manuscript; Dr Devillard managed the clinical case and critically reviewed and revised the manuscript; and all authors approved the final manuscript as submitted.

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the pediatric neurology department of the CHU of Bordeaux after episodes of tongue biting, increasing in both intensity and frequency, despite treatment with hydroxyzine, morphine, and acetaminophen. C.V. was whining, reacting to stimulation (noise, contact) with shouts and tongue biting. Her consciousness was altered. Many repeated episodes of tachycardia (up to 170 beats per minute) followed by sudden flexion of upper limb with clenched fists on her thorax, leg extension for a few seconds, shouting with tongue biting, and persistent trismus suggested the occurrence of complex partial seizures. Between the crises, there was no return to usual consciousness. Brain MRI showed a previously known right-sided stroke in the basal ganglia. Lesch–Nyhan syndrome was suspected but ruled out after tests revealed normal uric acid in blood and urine. Clonazepam was introduced at 0.05 mg then increased to 0.1 mg/kg per day, with delayed and partial efficacy. Because of a major episode of bleeding mouth, and with the goal of controlling dystonia by introducing high levels of analgesia and sedation, C.V. was transferred to the PICU of the CHU of Bordeaux for monitoring. During her hospitalization in PICU from April 15 to April 17, oxcarbazepine was introduced to treat choreodystonic movements, and clonazepam was continued. Despite the intensity of the treatment, chewing and biting episodes recurred, causing significant injuries, including a half split of the tongue (Fig 1, white star).

After a first multidisciplinary meeting of odontologists, pediatricians intensivists, and pediatric neurologists, on April 17 her tongue was sutured (Fig 2A) and mandibular central incisors were extracted in the operating room, under general anesthesia. A postoperative transfer to the PICU was necessary to maintain sedation and to prevent additional tongue biting. She was intubated and under mechanical ventilation. Hemoglobin was 6 g/dL. She received a blood transfusion. Sedation was achieved with sufentanil and midazolam, and muscular blockade was added for a few hours.

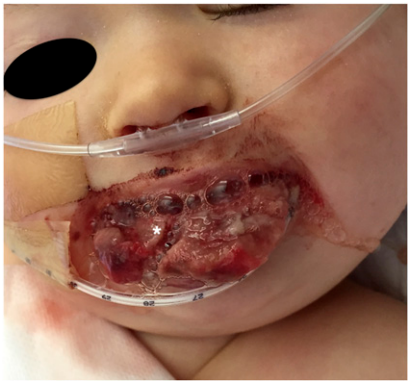

FIGURE 1
Severe injuries of the tongue.

A second multidisciplinary meeting was held on April 22 to limit

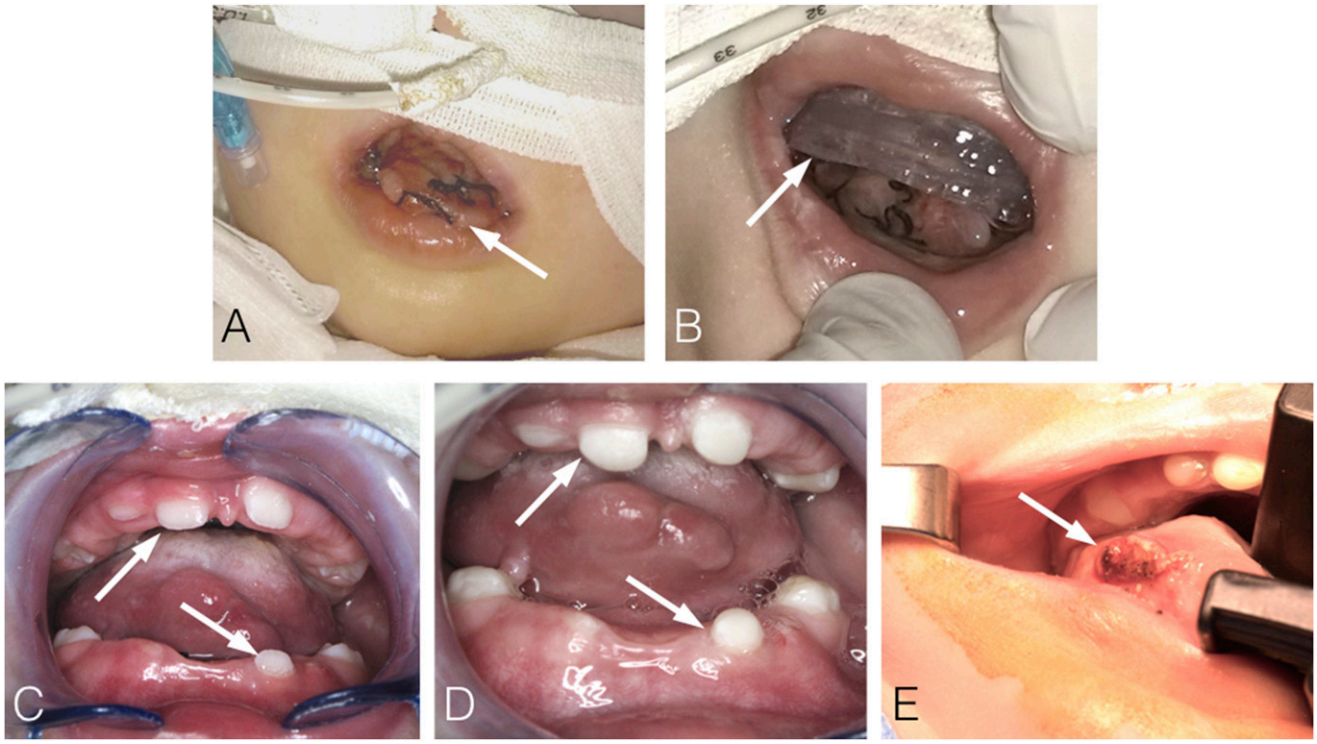

FIGURE 2
Progressive oral management. A, Tongue was sutured (white arrow). B, Tongue protection with bite guard (white arrow). C, Sharp-shaped teeth before composite applications (white arrows). D, Round-shaped teeth after composite applications (white arrows). E, Partial glossectomy with a diode laser (white arrow).

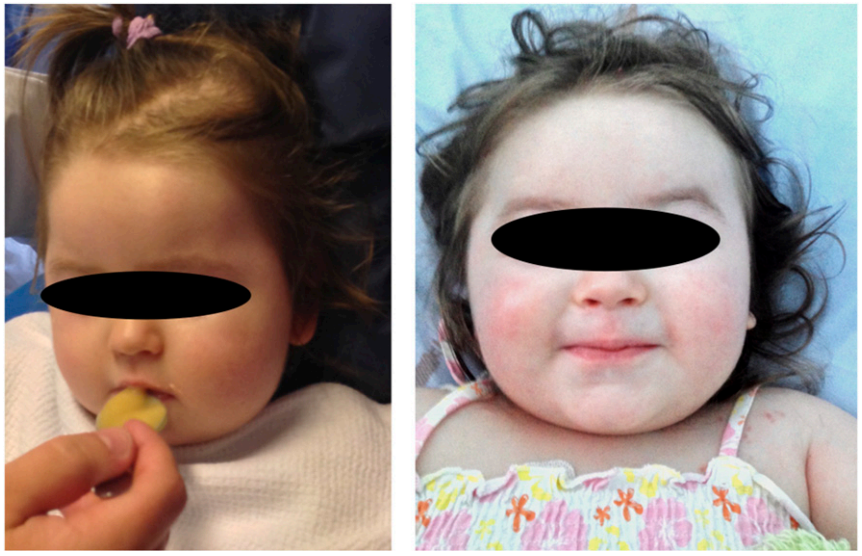

FIGURE 3
Patient fully awake and eating, with no disfigurement.

recurrence and protect the tongue from oromandibular dystonic movements (ODM) when the patient regained consciousness. A bite guard (Fig 2B, white arrow) was applied to protect the patient's tongue, and an antidystonic drug (trihexyphenidyl hydrochloride [Artane]) was introduced. On April 30, recurrence of choreodystonic movements was noted after curare discontinuation. First, levodopa (≤3 mg/kg per day) was combined with trihexyphenidyl hydrochloride, then levodopa was discontinued and tetrabenazine (Xenazine) was combined with Artane. On May 11, a third multidisciplinary meeting discussed the patient's treatment, and the following conclusions were reached:

- Oral management: Apply composites to smooth tooth surfaces (Fig 2 C and D) and avoid dental extractions because of risks of facial malformation during development.
- Neurologic management: Maintain sedation, analgesia, and muscular blockade with mechanical ventilation. Stop Artane because of its lack of efficacy, and increase the dosage of Xenazine (≤5.8 mg/kg per day).
- Discuss the possibility of injecting botulinum toxin in case of refractory ODM with persistent tongue damage.

An emergency fourth multidisciplinary meeting was organized after her general status worsened, and on May 22 it was decided to inject botulinum toxin into the masseters and orbicularis oris. Neuromuscular blockade (NMB) was discontinued on May 26. No resurgence of dystonic movements was noticed after the injections. Tooth eruption was checked regularly, and composites were added after the eruption. The bite guard was well tolerated. Sedation and analgesia were tapered slowly to prevent withdrawal syndrome. On May 28, a partial glossectomy (Fig 2E, white arrow) was performed under general anesthesia with a diode laser to remove damaged tissue and prevent excessive bleeding. Some teeth seemed to be mobile. We removed the bite guard and did not notice new soft tissue injuries. On June 5, the fifth and final multidisciplinary meeting took place. We discussed, a posteriori, the efficacy of the botulinum toxin injections and the need to plan a gastrostomy to encourage enteral feeding and promote autonomy. No dental extractions were necessary, and the bite guard was readjusted to accommodate newly erupting teeth. Gastrostomy was performed on June 16. C.V. was extubated on June 14. She was then fully awake. Oral alimentation was reintroduced progressively, and no disfigurement was observed (Fig 3).

DISCUSSION

We report a rare case of automutilation secondary to ODM in a young girl. In this case, tongue and lip self-injuries were so severe that she remained in the PICU with NMB and strong sedation for almost 4 weeks. To our knowledge, this is the first description of using resin composite on sharp temporary teeth to prevent tongue damage in an infant. We highlight the use of bite guard and botulinum toxin for this indication. We suggest contacting odontologists early to provide multidisciplinary management of these children.

Childhood-onset dystonia is not uncommon, and it is a challenge to physicians, necessitating management by a multidisciplinary team and close attention to dosage and side effects.[1]

Oromandibular dystonia is a specific form of dystonia characterized by involuntary, action-induced tonic or clonic spasms of the masticatory, lingual, and pharyngeal musculature. Symptoms include bruxism, dysphagia, soft tissue trauma, and temporomandibular joint subluxation.[2] The most common injuries are bites, located mostly on the tongue and the lower lip,[3] with sometimes severe injury, infection, loss of tissue, and subsequent scarring,[4] especially when repeated. If drug (levodopa, Xenazine, and Artane) use is typically

recommended for generalized choreodystonic manifestations, botulinum toxin injection should be discussed in focal cases.[1]

Patients with cerebral palsy often present with bites resulting from interposition of the tongue or lips between dental arches and self-injuries.[5] The risk of bruxism is increased by certain lesions of the central nervous system and exacerbated during intense emotions such as pleasure or pain.[5] Patients occasionally develop status dystonicus, a life-threatening condition characterized by increasingly frequent or continuous severe episodes of generalized dystonic spasms (contractions) necessitating urgent hospital management.[6,7] Several medications are reported to trigger status dystonicus: dopamine receptor blockers (pimozide and haloperidol), metoclopramide, and clonazepam.[7] To break the status dystonicus, continuous intravenous midazolam is usually chosen because of its muscle relaxant effect, rapid onset, and short half-life.[7] Strong sedation and muscle relaxation are the measures most likely to achieve prompt resolution of dystonic spasms.[7]

In the case of C.V., an important point was the serial misdiagnosis caused by the initial clinical presentation mimicking seizures. In the context of prenatal stroke and results of the MRI, we treated this case as a status epilepticus. Odontologists intervened early to suture the tongue and provide specialized expertise regarding the oral lesions. Odontologists recommended installing of a bite guard, a system that has shown efficacy in self-injuries due to other neurologic disorders such as Lesch–Nyhan syndrome.[8] The first attempt to discontinue NMB involved applying resin composites to the sharp temporary teeth. This noninvasive technique eliminated the sources of cuts and was successful for 8 days.

To our knowledge, no study has been published about composites preventing oral self-injury. A worsening of the general condition led us to use Botox injections. Botox injections are used for children with cerebral palsy,[9,10] posttraumatic oromandibular dystonia,[11] and secondary dystonia self-mutilation.[12] In this case, because of the young age of the patient, the use of botulinum toxins would be contraindicated but has been discussed in depth.

Botox infiltrations were administered by an experienced dental surgeon with the aid of sonography and electrical stimulation.[9] Infiltrations were made in the masseters and orbicularis oris. Botulinum toxin infiltration was our last resort as the patient's condition worsened. Her prognosis was uncertain. The infiltrations enabled us to withdraw NMB and sedation without the occurrence of new injuries. The antidystonic medication was then adapted to the generalized dystonia without interference of sedation. New eruptions and the efficacy of infiltrations were reevaluated after 3 and 6 months. No abnormal muscle contractions were observed.

The strong collaboration between the various therapists and the progressive oral therapies were critical to the success of the treatment. In the treatment of focal dystonia associated with severe injuries, botulinum toxin injection should be discussed much earlier.

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ABBREVIATIONS

CHU: University Hospital Center
NMB: neuromuscular blockade
ODM: oromandibular dystonic movements

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