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Case Report

# Bell's palsy in a pediatric patient with hyper IgM syndrome and severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2)

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## Abstract

Bell's palsy is an acute facial paralysis with known association to viral infections. We describe a medically complex 6-year-old male with hyper IgM syndrome who presented with unilateral facial droop and positive SARS-CoV-2 RT-PCR. This is the first reported pediatric case of Bell's palsy in the setting of SARS-CoV-2 infection.

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## 1. Introduction

Bell's palsy is an acute, unilateral facial paralysis characterized by lower motor neuron dysfunction of the facial nerve [1]. The condition occurs more frequently in adults but is also seen in pediatric patients. The condition is typically self-limiting, and symptoms generally resolve within weeks to months. In a minority of cases, however, it can result in longer-term facial muscle weakness or in sequelae such as ocular dryness, lagophthalmos, exposure keratopathy, or swallowing dysfunction [2]. The condition often resolves without treatment, however antiviral and corticosteroid management is typically utilized, especially if diagnosed within 72 h of symptom onset [3,4].

A broad array of other etiologies for Bell's palsy have been described including traumatic, inflammatory, and infectious processes [5–8]. In particular, the role of viral infection in association with the condition has been reported with herpetic viral infections observed as a frequent cause [5,6]. There are limited reports of an association between severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) infection and Bell's palsy in adults but this seems to be rare with only two cases reports at the time of this report. [9,10] Bell's palsy has never been reported in a child with SARS-CoV-2 seropositivity. Herein, we report the first case of a pediatric patient presenting with acute onset Bell's Palsy in the setting of SARS-CoV-2 infection.

## 2. Case report

A 6-year-old male with history of prematurity (born at 30 weeks gestation), failure to thrive, chromosome

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17 and 19 deletions, submucosal cleft palate, surgically repaired atrial and ventricular septal defects, agammaglobulinemia with hyper IgM, hypospadias, asthma, moderate obstructive sleep apnea, and gastrostomy tube feeding presented to the pediatric emergency room with one day history of right sided facial droop. Parents reported an asymmetric smile, drooling, and inability to fully close the right eye. They denied fever or vomiting at home and felt the patient was otherwise behaving at baseline. There was no history of trauma, sick contacts, or recent travel. At baseline, the patient received intravenous immunoglobulin (IVIG) infusion every three weeks for his agammaglobulinemia but was on no other pharmacotherapy.

On evaluation, the patient was tachycardic to 116 beats/minute but otherwise afebrile, normotensive and well-appearing. Physical examination and neurologic examination were unremarkable except for a right-sided facial palsy with inability to close the right eye and right sided mouth droop with drooling (House-Brackmann grade: IV). Laboratory testing was significant for an elevated white count at 11.1 K/uL. SARS-CoV-2 Reverse transcription polymerase chain reaction (RT-PCR) nasopharyngeal swab was positive. Herpes Simplex Virus (HSV-1, HSV-2) and Varicella Zoster Virus (VZV) PCR were negative. Of note, the patient had tested negative for SARS-CoV2 during a prior admission two weeks earlier. There were no known contacts or family members with SARS-CoV-2 infections. A respiratory viral panel ten days prior to admission was negative for adenovirus, coronavirus HKU1/229E/NL63E/OC43, metapneumovirus, rhinovirus, enterovirus, influenza A and B, parainfluenza 1–4, and respiratory syncytial virus.

The patient was started on intravenous acyclovir 150 mg every 8 h and received his scheduled dose of IVIG infusion. Lubricating eye drops were initiated as well for corneal protection. During admission, a magnetic resonance imaging (MRI) was attempted but could not be completed due to patient movement. The patient was not sedated for the imaging study due to the SARS-CoV2 positive status.

Once stable, the patient was discharged on a five-day course of prednisolone and acyclovir. At follow-up in three weeks, the symptoms had improved (House-Brackmann grade: I).

### 3. Discussion

To our knowledge, this is the first report of an association between Bell's palsy and SARS-CoV-2 in a pediatric patient. Few cases of Bell's palsy in the setting of SARS-CoV2 infection have been reported in adults and appear to be very infrequent [9,10]. In these cases, facial nerve palsy was noted in the setting of fevers and viral symptoms and workup for other typical causes

of Bell's palsy were negative. Of note, our patient presented with no reported history of viral symptoms nor pulmonary complaints and the facial droop was the only noted symptom at presentation. The timeline of our patient's negative SARS-CoV2 RT-PCR during hospitalization two weeks prior to presentation coupled with the negative VZV and HSV PCR tests is particularly suspicious of causative infection with SARS-CoV-2. Unlike prior reports of Bell's palsy [9,10], the patient was definitively SARS-CoV-2 negative just two weeks prior making the temporal relationship with the onset of symptoms difficult to ignore.

Although there is a well-established relationship between Bell's palsy and viral infection, this has tended to be associated with members of the Herpesviridae family. HSV-1 DNA has been detected in the *endo*-neural fluid of the facial nerve and around the geniculate nucleus in patients with Bell's palsy [11,12]. In particular, HSV and VZV infections have been most commonly described, however a broader array of viruses have been implicated in Bell's palsy including Epstein-Barr virus, Human herpesvirus 6 (HHV-6), human immunodeficiency virus (HIV), Hepatitis B, influenza, adenovirus, rubella, mumps, and coxsackievirus [13–18]. As with the adult cases, the possibility that the Bell's palsy was an unrelated occurrence that just presented coincidentally alongside the SARS-CoV2 infection cannot be ruled out.

From a therapeutic standpoint, the administration of steroid treatment was debated given the patient's underlying immunocompromised status. There are limited evidence-based guidelines for treatment of immunocompromised children with Bell's palsy. However, the use of steroids in the management of acute onset Bell's palsy has been more thoroughly discussed [19]. Given the severity of facial symptoms (House-Brackmann grade > IV), short time frame to presentation and the patient's ongoing IVIG treatment, it was felt that the patient's agammaglobulinemia was well enough controlled to warrant trial of steroid treatment. He was closely monitored after discharge and improved quite well on this regimen although as this is a singular case, further extrapolation is not possible.

Our patient's history of hyper-IgM syndrome may complicate his response to a SARS-CoV-2 infection. Children with hyper-IgM syndrome are known to have immunodeficiency due to shifted immunoglobulin production towards IgM and away from other isotypes such as IgG and IgA. However, their immunodeficiency is often due to an inability to yield long-term immune protection such as with the development of IgG antibodies, and thus, for an initial infection with SARS-CoV-2, this may be of less clinical importance as compliment cascades and opsonization of antigens are preserved in this population. Hyper IgM syndrome patients also have

defective cellular immunity which can increase susceptibility to viral infections.

The manifestations of SARS-CoV2 virus in children are still being understood. In pediatric patients, respiratory illness, leukopenia, thrombocytopenia, myocarditis, interstitial pneumonia, Kawasaki-like illness, and vasculitis have all been reported [20]. Our case highlights the possibility of atypical, neurologic, presentations of the virus in pediatric patients. Further research is needed to fully understand the role of SARS-CoV2 infection in patients with Bell's palsy, however in the context of the current coronavirus disease 2019 (COVID-19) pandemic, we recommend practitioners consider it as a potential atypical presenting sign of the infection.

### Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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