Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Pediatrics

Hopkins Syndrome after an Asthma Exacerbation: A Case Report

Hafsa Benquassmi[1*], Ahlam Mosadik[1] Alae Elkoraichi[1], Salma Ech-Cherif Kettani[1], Aziza Bentalha[1]

[1]Pediatric Intensive Care Anesthesia Department, Pediatric Hospital of Rabat, IBN SINA University Hospital Rabat, Morocco

DOI: 10.36347/sjmcr.2024.v12i05.085 | **Received:** 16.04.2024 | **Accepted:** 23.05.2024 | **Published:** 27.05.2024

***Corresponding author:** Hafsa Benquassmi
Pediatric Intensive Care Anesthesia Department, Pediatric Hospital of Rabat, IBN SINA University Hospital Rabat, Morocco

Abstract **Case Report**

Hopkins Syndrome, also known as Hopkin-McCarthy Syndrome or post-asthmatic paralysis, is a rare condition characterized by the onset of acute flaccid paralysis in a child following an asthma exacerbation. Although the underlying mechanisms are not fully understood, this syndrome is believed to be a form of myelopathy associated with asthma. We report a case of Hopkins syndrome occurring after an episode of asthma exacerbation in a 2-year and 8-month-old boy.

Keywords: Hopkins Syndrome, asthma exacerbation, rare disease, radiculopathy.

INTRODUCTION

Hopkins syndrome (HS) is an acute flaccid paralysis occurring a few days to weeks after an asthma exacerbation. It was first reported in 1974 in Australia [1] and only around forty cases have been reported. Therefore, it is essential to take notice of this case report that sheds light on this poorly documented condition.

CASE DESCRIPTION

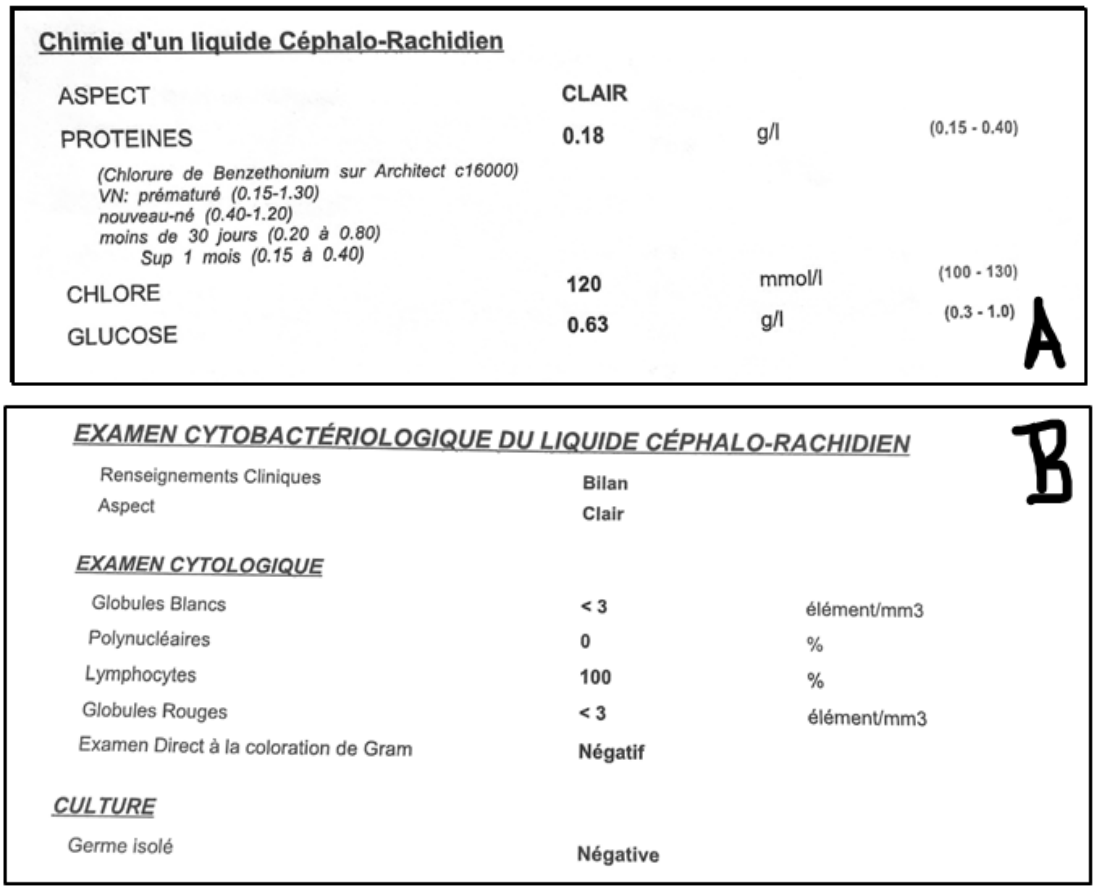
We report the case of a 2-year and 8-month-old boy who had been immunized according to the national program program, with a history of asthma since being 9 months old He had used a salbutamol inhaler, and his mother had a history of atopy. He was admitted to the intensive care unit for severe asthma exacerbation, and treated with nebulized salbutamol (150µg/kg/nebulization) + Ipratropium bromide (0.25 mg/Kg/8h) in nebulization with intravenous magnesium sulfate at a rate of 30 mg/kg, corticosteroid at 2 mg/kg and intravenous beta dose adrenaline given the non-improvement. Our patient also benefited from antibiotic therapy based on C3G and aminoglycoside in the absence of amoxicillin for suspected community-acquired pneumonia including a fever of 39.2°, purulent secretions, a right basal pulmonary focus on chest x-ray, His complete blood count was as follows: 28,700 /mm3 white cells/mm^3 with 81% neutrophils. C-reactive protein (CRP) was 25 mg/l, and procalcitonin was 1.25 ng/ml. Arterial blood gas analysis revealed respiratory failure and respiratory acidosis (pH: 7.30, PaCO2: 64 mm Hg, PaO2: 66 mm Hg, bicarbonate: 26) under a high concentration oxygen mask (15 L). At PDP: Bacillus Gram-negative with culture-negative, multiplex PCR of nasopharyngeal was negative. The patient didn't show any signs of improvement and required mechanical ventilation for 4 days, the sedation was necessary. After normalizing the arterial gas, apyrexia, absence of wheezing, and radiographic signs of pneumopathy, the sedation was stopped and the patient was extubated.

On the 5th day, the child presented muscular weakness in all four limbs but more marked in the lower limbs. On clinical examination: contact was present, flaccid quadriparesis, deep tendon reflexes (ROT) were present but weak, plantar skin reflex in flexion, sensitivity was preserved and the cranial nerves were intact.

Several diagnoses have been mentioned:

- Cerebral hypoxia: the patient has never experienced profound hypoxia.
- Critical Illness Polyneuropathy (despite the introduction of corticosteroids, curare, and mechanical ventilation) but eliminated after a negative muscle biopsy.
- Guillain-Barre Syndrome: An albumin cytological dissociation was not noted at CSF examination (Figure 1). However, the EMG suggested a motor axonal poly neuropathic attack involving the territory of the L4 L5 roots bilaterally which could fall within the framework of motor form syndrome of Guillain barre (Figure 2). The decision was to initiate intravenous immunoglobulin therapy at a rate of 0.4 g/kg/day for 5 days, along with motor

physiotherapy of both lower limbs. The evolution was marked by a clear improvement with total recovery of his motor skills then transferred to the neuropediatric department for further treatment.



Chimie d'un liquide Céphalo-Rachidien

ASPECT	CLAIR		
PROTEINES	0.18	g/l	(0.15 - 0.40)
(Chlorure de Benzethonium sur Architect c16000) *VN: prématuré (0.15-1.30)* *nouveau-né (0.40-1.20)* *moins de 30 jours (0.20 à 0.80)* *Sup 1 mois (0.15 à 0.40)*			
CHLORE	120	mmol/l	(100 - 130)
GLUCOSE	0.63	g/l	(0.3 - 1.0)

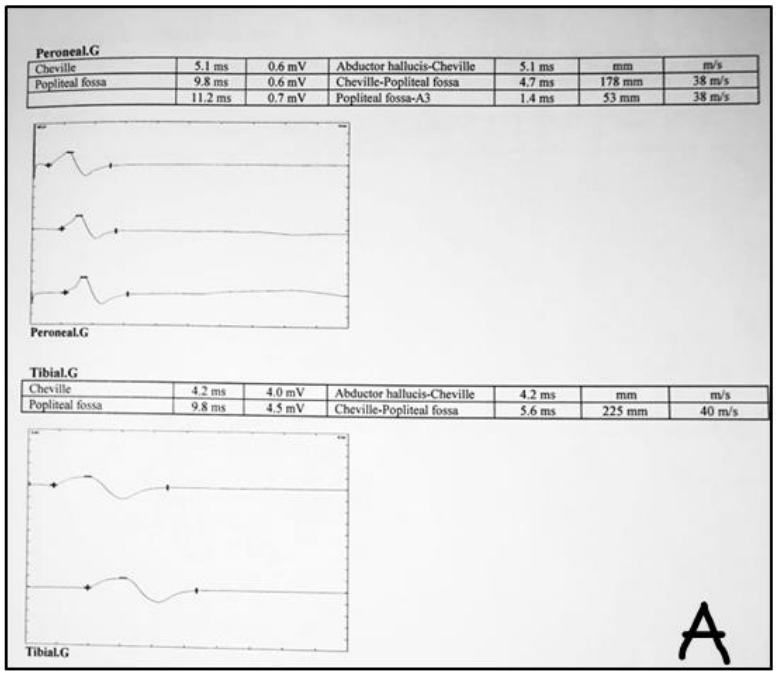
A

EXAMEN CYTOBACTÉRIOLOGIQUE DU LIQUIDE CÉPHALO-RACHIDIEN

B

Renseignements Cliniques	Bilan	
Aspect	Clair	
EXAMEN CYTOLOGIQUE		
Globules Blancs	< 3	élément/mm3
Polynucléaires	0	%
Lymphocytes	100	%
Globules Rouges	< 3	élément/mm3
Examen Direct à la coloration de Gram	Négatif	
CULTURE		
Germe isolé	Négative	

Figure 1: Cerebrospinal fluid analysis (CSF). A: chemicals in the CSF. B: cytological and bacteriological examination of CSF



Peroneal.G

Cheville	5.1 ms	0.6 mV	Abductor hallucis-Cheville	5.1 ms	mm	m/s
Popliteal fossa	9.8 ms	0.6 mV	Cheville-Popliteal fossa	4.7 ms	178 mm	38 m/s
	11.2 ms	0.7 mV	Popliteal fossa-A3	1.4 ms	53 mm	38 m/s

Peroneal.G

Tibial.G

Cheville	4.2 ms	4.0 mV	Abductor hallucis-Cheville	4.2 ms	mm	m/s
Popliteal fossa	9.8 ms	4.5 mV	Cheville-Popliteal fossa	5.6 ms	225 mm	40 m/s

Tibial.G

A

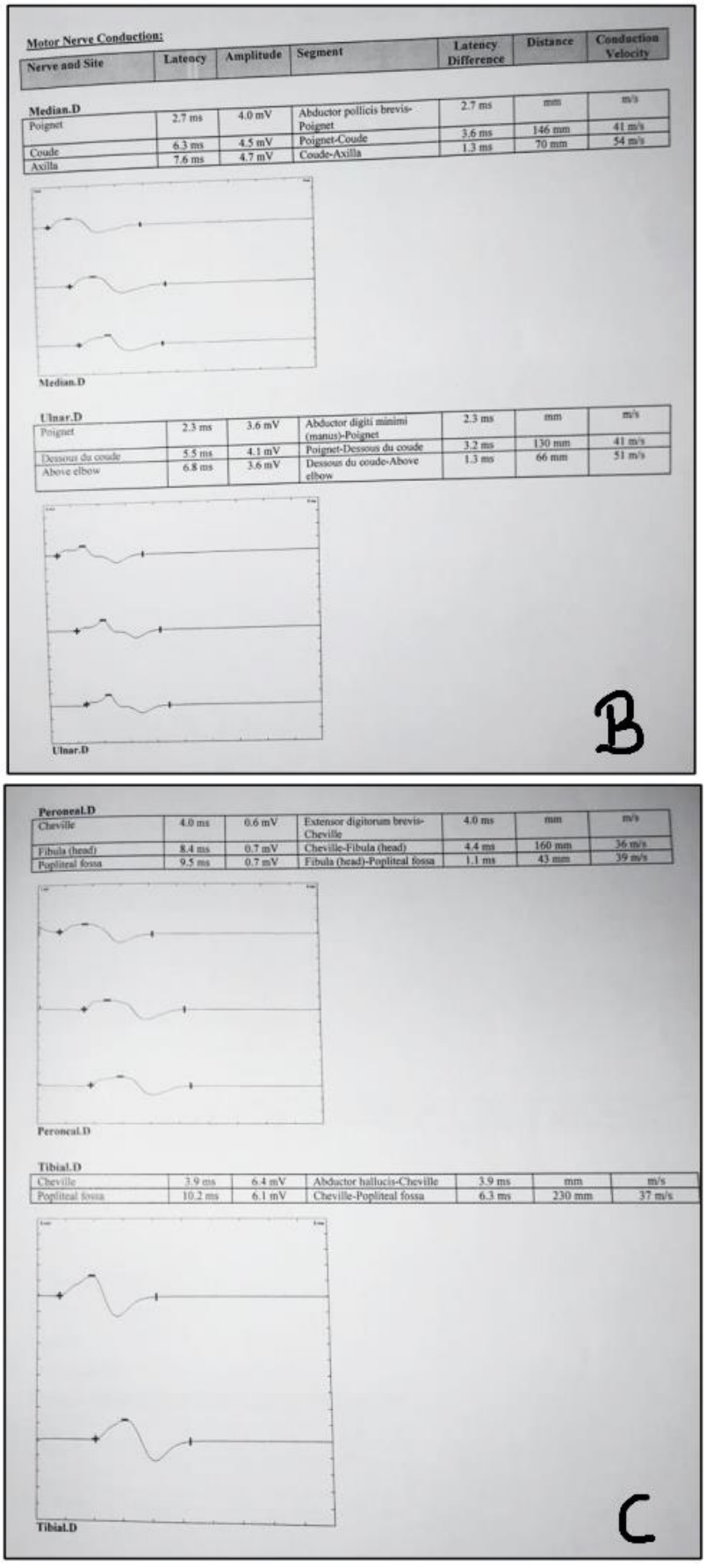


Figure 2: Electromyography exam. A-B-C: motor nerve conduction

DISCUSSION

Due to its rarity, Hopkins syndrome is poorly documented in the medical literature. Published cases are generally limited to isolated case reports and small case series.

Although the pathogenesis of HS is not understood, many explanations have been considered infectious and immune mechanisms [2, 3]. Indeed, this syndrome could be a form of autoimmune response where severe inflammation linked to asthma exacerbation could induce an immune response which, by mistake, targets the anterior horn cells of the spinal cord [4]. Involvement of the pyramidal tracts and posterior horns was also found on spinal cord MRI [5].

Thus, it has been reported that lymphocytes of asthmatic patients express more cytokines upon stimulation by allergens [6], most often in children suffering from atopic asthma. Still, the most likely hypothesis is that this weakness could be caused by a neurotropic virus (enterovirus D68) [7], which would have triggered an asthma exacerbation and could affect cells in the anterior horn of the spinal cord or peripheral nerves. This virus was found in two teams:

- An American, from cerebrospinal fluid samples or stool from pneumonia patients.
- A Japanese, 3/205 children with enterovirus D68 pneumonia (detected at PDP) presented acute flaccid paralysis. In our patient, nasopharyngeal and CSF PCR multiplex did not reveal any viruses.

The diagnosis of Hopkins syndrome is clinical, it is often a diagnosis of exclusion. Generally, this disease manifests a few days to a few weeks after an asthma exacerbation. For our patient, the time was 5 days. It presents as an acute flaccid paralysis that can affect one or more limbs and develop gradually into severe muscular atrophy which in most cases progresses to severe muscle atrophy in the absence of physiotherapy.

In our patient, the Electromyography exam mentioned a poly neuropathic motor axonal attack affecting the territory of the L4 L5 roots bilaterally which could fall within the framework of the motor form of Guillain-Barre syndrome. And no albumin cytological dissociation has been found. The spinal MRI was normal.

Hopkins syndrome is frequently associated with epilepsy. This was reported in the patients of Pitt and Hopkins (1978) and Singh (1993). In our patient, no clinical or electrical epileptic seizures were reported.

Based on previous studies, there is no consensus on treatment [8]. However, the recommendations are made on a case-by-case basis. The treatment proposed was intravenous immunoglobulins 0.4 g/Kg/day for 5 consecutive days followed by methylprednisolone at a rate of 30 mg/kg/day for 3 days, which was started 23 days after the onset of symptoms and improvement was after 3 weeks [9]. In our patient, intravenous immunoglobulins were administered 24 hours after the onset of the symptoms at a dose of 0.4g/kg/day for 5 days. The patient was already on corticosteroids at a rate of 2m/kg/day for his exacerbation asthma.

CONCLUSION

Hopkins syndrome is a rare entity, which remains a diagnosis of elimination. However, further studies are needed to clarify the etiology of this unusual syndrome. Early treatment is necessary for full recovery.

Conflicts of interest: The authors declare no conflict of interest.

REFERENCES

1. Mekmangkonthong, A., Khusiwilai, K., & Paticheep, S. (2022). Acute Flaccid Monoplegia After an Asthmatic Attack. *Asian Medical Journal and Alternative Medicine*, *22*(2), 153-156.
2. Gateau, K. L., David, H., & Lowe, C. G. (2019). Hopkins Syndrome: Post Flaccid Paralysis After an Asthma Exacerbation. *Pediatric Emergency Care*, *35*(10), e190-e191.
3. Gateau, K. L., David, H., & Lowe, C. G. (2019). Hopkins Syndrome: Post Flaccid Paralysis After an Asthma Exacerbation. *Pediatric Emergency Care*, *35*(10), e190-e191.
4. Arakawa, H., Hamasaki, Y., Kohno, Y., & Ebisawa, M. (2017). Allergology international Japanese guidelines for childhood asthma 2017. *Allergol Int*, *66*, 190-204.
5. Hixon, A. M., Clarke, P., & Tyler, K. L. (2017). Evaluating treatment efficacy in a mouse model of enterovirus D68–associated paralytic myelitis. *The Journal of infectious diseases*, *216*(10), 1245-1253.
6. Maloney, J. A., Mirsky, D. M., Messacar, K., Dominguez, S. R., Schreiner, T., & Stence, N. V. (2015). MRI findings in children with acute flaccid paralysis and cranial nerve dysfunction occurring during the 2014 enterovirus D68 outbreak. *American Journal of Neuroradiology*, *36*(2), 245-250.
7. Cantarín-Extremera, V., González-Gutiérrez-Solana, L., Ramírez-Orellana, M., López-Marín, L., Duat-Rodríguez, A., & Ruíz-Falcó-Rojas, M. L. (2012). Immune-mediated mechanisms in the pathogenesis of Hopkins syndrome. *Pediatric neurology*, *47*(5), 373-374.
8. Piero, P., Longo, M. R., Scalia, F., Polosa, R., Kira, J. I., & Falsaperla, R. (2010). Recurrent Hopkin's syndrome: A case report and review of the literature. *Journal of the neurological sciences*, *297*(1-2), 89-91.
9. Kim, Y. M., Orvedahl, A., Morris, S., Schmidt, R., & Mar, S. (2017). A 12-year-old girl with encephalopathy and acute flaccid paralysis: A neuropathological correlation and cohort review. *Pediatric Neurology*, *66*, 5-11.