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Acute infectious chorea in children: A study of 17 cases

Khalifa Ababacar Mbaye, Rokhaya Diagne, Serigne Saliou Mbacké, Ahmadou Bamba Mbodji, Papa Souleye Sow, Fatou Kane, Moustapha Ndiaye

ABSTRACT

Introduction: The objective of our study was to characterize the epidemiological, clinical, paraclinical, etiological, therapeutic, and evolutionary aspects of acute infectious chorea in children at the Albert Royer National Children's Hospital.

Methods: This was a retrospective descriptive study from January 2005 to January 2020, which took place in the pediatric neurology unit of the Albert Royer National Children's Hospital. All patients presenting with acute choreic syndrome in an infectious context and whose age was less than or equal to 16 years were included in our study. Patients with incomplete records were excluded.

Results: In 15 years, we collected 17 patients including 9 girls. The average age was 8.41 years. Clinically, chorea was generalized in 14 patients (82.35%) and localized in 3 patients (17.64%). Brain magnetic resonance imaging (MRI) showed panencephalitis in two patients and bithalamic and cortical hypersignals in one patient. Twelve patients were diagnosed with Sydenham's chorea (SC) (70.58%), four patients had chorea following herpes simplex infection and one patient had chorea secondary to enterovirus. Haloperidol was the most used treatment. The evolution was favorable for all children diagnosed with SC compared to other causes of chorea (viral chorea).

Conclusion: Chorea is a rare pathology in children and is dominated in our context by SC.

Keywords: Children, Chorea, Infectious, Senegal

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INTRODUCTION

Chorea is one of the hyperkinetic syndromes of abnormal movements with a prevalence of 5% of all abnormal movements in children [1]. There are many etiologies for chorea. In Africa, most studies focus on Sydenham's chorea and this is explained by the difficulty in finding the etiology [2, 3]. The objective of our study was to characterize the epidemiological, clinical, paraclinical, etiological, therapeutic, and evolutionary aspects of acute chorea of infectious origin in children at the Albert Royer National Children's Hospital.

MATERIALS AND METHODS

This was a retrospective descriptive study from January 2005 to January 2020 which took place at the pediatric neurology unit of the Albert Royer National Children's Hospital. All patients with acute choreic syndrome who were seen in a neurology consultation and whose age was less than or equal to 16 years were included in our study. Seven patients with incomplete records (incomplete

history of disease, missing clinical data, biological results, and brain imaging) were excluded.

The study of the follow-up files of the patients gave us information on the following characteristics:

- Socio-demographics (age, sex)
- Clinical aspects (type of chorea, mode of onset, circumstances of onset, neurological and extraneurological signs)
- The results of brain imaging [computed tomography (CT) and/or magnetic resonance imaging (MRI)] and other paraclinical data for etiological purposes (analyses: cytological, bacteriological, viral, and chemical of cerebrospinal fluid). Blood count, Antistreptolysin O (ASLO), C-reactive protein (CRP), and sedimentation rate (SR) were routinely requested. Cardiac ultrasound and electrocardiogram were only requested in children diagnosed with Sydenham's chorea. Lumbar puncture was only performed in children whose clinical picture did not suggest Sydenham's chorea. Statistical analysis of our data was performed using Excel 2016. Univariate analyses were performed to calculate frequencies and means.

RESULTS

In 15 years, we collected 17 patients, 9 of whom were girls. The average age was 8.41 years. The age group 6–10 years was the most represented with a percentage of 47.05% (see Table 1). The medical history was a notion of angina in the month preceding the chorea (12 patients), acute gastroenteritis (2 patients), influenza syndrome (2 patients), and allegation fever (1 patient).

Clinically, the onset of chorea was insidious in all patients. They were generalized in 14 patients (83.33%) and localized in 3 patients (16.66%). The localized forms were hemichorea (2 patients) and monochorea (1 patient). The associated neurological signs were hypotonia, which was found in all patients, dysarthria (3 patients), athetosis (4 patients), dystonia (1 patient), tonic-clinical generalized seizures (2 patients), and hemiparesis (1 patient). The extra-neurological signs were polyarthralgia, which were found in 4 patients.

Biologically, we noted a non-specific biological inflammatory syndrome in 8 patients (elevated CRP and SR) and positive ASLO (greater than 400) in 7 patients. Cerebral CT scans were performed in 15 patients and did not reveal any abnormalities. Brain MRI was performed in 7 patients and showed panencephalitis in 2 patients and bithalamic and cortical hypersignals in 1 patient. The electroencephalogrphy (EEG) revealed diffuse slowing in 2 patients.

Etiologically, 12 children had Sydenham's chorea (SC) and 5 children had chorea secondary to a viral infection [herpes simplex virus (4 patients) and enterovirus (1 patient)]. All patients diagnosed with Sydenham's chorea were treated with a neuroleptic (haloperidol, 0.2 mg/kg/d) for a mean duration of 4.08 months, with extremes of 1 to 12 months. Three patients received clonazepam (0.05 mg/kg/d) in addition to haloperidol. Eight patients received prednisone-based corticosteroids (2mg/kg/d) for 12 weeks. All children diagnosed with SC received penicillin-based antibiotic therapy as a matter of course. The three children diagnosed with chorea secondary to herpes simplex virus infection received intravenous methylprednisolone (1g/1.73 m^2) for five days, acyclovir injection (10 mg/kg/8 h) for 21 days and haloperidol (0.2 mg/kg/d) for 2–9 months depending on the patient. The patient diagnosed with chorea secondary to enterovirus had received methylprednisolone (1g/1.73 m^2) intravenously for five days, then prednisone (1 mg/kg/d) orally and haloperidol (0.2 mg/kg/d) for 10 months.

The evolution was favorable in 90.66% of our patients diagnosed with SC with remission times ranging from 1 to 12 months and a mean of 4.45 months. One patient had a relapse after early discontinuation of treatment. Children diagnosed with chorea secondary to herpes simplex virus infection had a less favorable outcome with one death, one lost to follow-up and one child with persistent symptoms.

The clinical, paraclinical, and therapeutic characteristics of each patient are summarized in Tables 2 and 3.

Table 1: Distribution of patients according to age groups

Age range (years)	Number (N)	Percentage (%)
(0–5)	4	23.52
(6–10)	8	47.05
(11–15)	5	29.41
Total	17	100

Table 2: Summary table of children diagnosed with Sydenham's chorea

Patients	Age	Sex	Years	Type	Extra-neurological signs	ASLO ≥400 IU/mL	Brain imaging	Treatment	Evolution (months)
1	15	M	2006	GC	MI+PA	(+)	CT:N	HL+CL+ cortico	F (12)
2	13	F	2006	GC	MI+PA	(+)	CT:N	HL+ cortico	F (6)
3	8	F	2009	HC	MI+AI	(–)	CT:N	HL+ cortico	F (4)
4	12	F	2009	GC	MI	(+)	CT:N	HL+ cortico	F (3)
5	10	M	2010	GC	MI	(+)	MRI:bi-thalamic hypersignal	HL+ cortico	F (2)
6	9	F	2010	GC	MI+ AI+PA	(+)	MRI:N	HL+ cortico	F (9)
7	7	F	2010	GC	(–)	(–)	–	HL	F (1)
8	9	M	2016	GC	MI	(+)	CT:N	HL+ cortico	F (7)
9	5	F	2016	HC	(–)	(–)	CT:N	HL	F (2)
10	8	M	2017	GC	(–)	(–)	CT:N	HL+CL	R
11	8	F	2017	GC	MI	(+)	–	HL+CL+ cortico	F (1)
12	11	M	2020	GC	PA	(–)	MRI:N	HL	F (2)

GC: generalized chorea; HC: hemichorea; PA: polyarthralgia; MI: mitral insufficiency; AI: aortic insufficiency; CT: computed tomography; MRI: magnetic resonance imaging; N: normal; HL: haloperidol; CL: clonazepam; cortico: corticosteroid; F: favorable; R: recurrence

Table 3: Summary table of patients diagnosed with other infectious choreas

Patients	Age	Sex	Type	History	Neurological signs	Brain imaging	Lumbar puncture	Treatment	Evolution
1	2	F	GC	Acute febrile gastroenteritis	Athetosis+ GTCS	MRI:panencephalitis	29L, P :0.32 ; G:0.49 Herpes simplex virus	HL+CL+C	Lost to view
2	3	M	GC	Influenza-like illness	Athetosis+ GTCS	MRI:N	42L, P :0.28 ; G:0.61 Herpes simplex virus	HL+CL+C	Deaths
3	3	M	GC	Acute febrile gastroenteritis	Athetosis+ GTCS	MRI:panencephalitis	123L, G:0.69 P:0.35; enterovirus	HL+CL+C	Favorable (10 months)
4	9	M	MC (RUM)	Allegation fever	Dystonia + HP	CT:N	G :0.58; P :0.79 Herpes simplex virus	HL+C	Persistence
5	11	F	GC	Influenza-like illness	Athetosis	CT:N	85L, P:0.41; G:0.72 Herpes simplex virus	HL+C	Favorable (9 months)

GC: generalized chorea; MC: monochorea; RUL: right upper limb; GTCS: generalized tonic-clonic seizures; HP: hemiparesis; CT: cerebral tomography; HL: haloperidol; CL: clonazepam; L: lymphorachia; P: proteinorachia; G: glycorachia.

DISCUSSION

In 15 years, we collected 17 patients of whom 12 (70.58%) had SC. It is the most common acquired chorea in children [4]. Its high prevalence in developing countries can be explained by poor hygiene and low socio-economic status. The improvement in living standards, the advent of antibiotics, and prophylactic measures had made the condition rare in developed countries [2]. A female predominance was found in our study in accordance with the literature [5, 6]. In our study, chorea was generalized in 83.33 % of patients and the localized forms (16.66%) are either to a hemibody (hemichorea) or to a limb segment (monochorea) [2, 3]. Associated signs are dominated by valvular disease. Cardiac involvement is the major prognostic element and the main cause of the severity of the disease; its frequency varies from 20% to 35% [7].

The symptomatic treatment of SC is nowadays multiple. Some drugs such as the neuroleptics

(haloperidol, pimozide, and chlorpromazine) have always shown their efficacy with a clear superiority of haloperidol over the latter; however, the existence of side effects is more noted in patients on haloperidol [8]. Antiepileptic drugs such as carbamazepine and sodium valproate are also used in the symptomatic treatment of SC with a superiority of antiepileptic drugs over neuroleptics due to their lower risk of side effects. In our study the patients were all on haloperidol with good clinical improvement [8, 9]. All our patients received penicillin-based antibiotic therapy. A 10-day course or a single intramuscular injection of penicillin is recommended even in the absence of signs of evolving streptococcal infection [10]. In our study, as in the literature, the evolution is usually favorable after treatment and a few rare cases of relapse or persistence of chorea have been described, most often due to non-compliance with treatment or the existence of another underlying cause [8].

In our study, infectious meningoencephalitis was the second most common cause of acute infectious chorea in children. The etiological research was difficult in our context for economic and technical reasons. Enterovirus and herpes simplex virus were the only virus found contrary to the literature where other germs such as, mycoplasma [11], HIV [12], tuberculosis [13] are found. Magnetic resonance imaging of the brain showed no specific lesions, but panencephalitis in 2 patients. These brain lesions are not always present or concomitant with the infection. The evolution of chorea after meningoencephalitis is usually favorable [14], which was contrary to our results where we noted two recoveries after 9 and 10 months of treatment. This is explained by the difficulty in diagnosis (delay and diagnostic means) and the limited therapeutic means in our context (immunoglobulin, plasma exchange) [14–16].

CONCLUSION

Chorea is an uncommon symptomatology in our context. In our study, Sydenham's chorea is the most represented form of acute infectious chorea in children. Treatment with haloperidol remains effective in the symptomatic treatment of chorea.

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Author Contributions

Khalifa Ababacar Mbaye – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Rokhaya Diagne – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.