Acute ataxia in paediatric emergency departments: a multicentre Italian study

Giacomo Garone,[1] Antonino Reale,[2] Nicola Vanacore,[3] Pasquale Parisi,[4] Claudia Bondone,[5] Agnese Suppiej,[6,7] Giacomo Brisca,[8] Lucia Calistri,[9] Duccio Maria Cordelli,[10] Salvatore Savasta,[11] Salvatore Grosso,[12] Fabio Midulla,[13] Raffaele Falsaperla,[14] Alberto Verrotti,[15] Elena Bozzola,[16] Cristina Vassia,[5] Liviana Da Dalt,[17] Rosario Maggiore,[8] Stefano Masi,[9] Lucia Maltoni,[10] Thomas Foiadelli,[11] Annalisa Rossetti,[12] Carla Greco,[13] Silvia Marino,[14] Claudia Di Paolantonio,[15] Laura Papetti,[18] Antonio Francesco Urbino,[5] Rossella Rossi,[2] Umberto Raucci[2]

For numbered affiliations see end of article.

Correspondence to
Dr Umberto Raucci, Emergency Department, Bambino Gesù Children Hospital, Rome 00165, Italy; umberto.raucci@opbg.net

GG and UR contributed equally.

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ABSTRACT

Objectives To evaluate the causes and management of acute ataxia (AA) in the paediatric emergency setting and to identify clinical features predictive of an underlying clinically urgent neurological pathology (CUNP).

Study design This is a retrospective medical chart analysis of children (1–18 years) attending to 11 paediatric emergency departments (EDs) for AA in an 8-year period. A logistic regression model was applied to identify clinical risk factors for CUNP.

Results 509 patients (mean age 5.8 years) were included (0.021% of all ED attendances). The most common cause of AA was acute postinfectious cerebellar ataxia (APCA, 33.6%). Brain tumours were the second most common cause (11.2%), followed by migraine-related disorders (9%). Nine out of the 14 variables tested showed an OR >1. Among them, meningeal and focal neurological signs, hyporeflexia and ophthalmoplegia were significantly associated with a higher risk of CUNP (OR=3–7.7, p<0.05). Similarly, the odds of an underlying CUNP were increased by 51% by each day from onset of ataxia (OR=1.5, CI 1.1 to 1.2). Conversely, a history of varicella-zoster virus infection and vertigo resulted in a significantly lower risk of CUNP (OR=0.1 and OR=0.5, respectively; p<0.05).

Conclusions The most frequent cause of AA is APCA, but CUNPs account for over a third of cases. Focal and meningeal signs, hyporeflexia and ophthalmoplegia, as well as longer duration of symptoms, are the most consistent 'red flags' of a severe underlying pathology. Other features with less robust association with CUNP, such as seizures or consciousness impairment, should be seriously taken into account during AA evaluation.

What is already known on this topic?

- Ataxia is a relatively uncommon neurological emergency in childhood.
- Acute postinfectious cerebellar ataxia and intoxications are the most common causes of acute paediatric ataxia.

What this study adds?

- In Italy, acute postinfectious cerebellar ataxia is the most common cause of acute ataxia in children, and varicella zoster is the most frequently involved pathogen.
- Brain tumours are the second most common cause of paediatric acute ataxia.
- In acute ataxia assessment, focal neurological or meningeal signs, hyporeflexia, ophthalmoplegia, seizures, and longer duration of symptom evolution are associated with higher risk of severe underlying pathology.

INTRODUCTION

Ataxia consists of impaired coordination of motor activity. Children with ataxia classically present a wide-based gait, truncal instability, tremor, dysarthria and nystagmus.[1–4] Acute ataxia (AA) in childhood poses a diagnostic dilemma because of the broad differential diagnosis. While the most common causes are benign, AA can be due to potentially disabling or life-threatening conditions, requiring early diagnosis and prompt intervention.[1] Only few studies have described the different conditions that can be encountered in children presenting with AA, but conclusions were divergent due to different recruitment settings, small sample size and heterogeneous study designs.[5–8]

The first aim of our study was to describe the demographic and clinical features of children presenting to the paediatric emergency department (PED) for AA in a large, multicentre cohort, investigating the underlying aetiologies and analysing the management in an emergency setting. Given the critical importance of identifying patients requiring urgent investigations, our second aim was to identify clinical features associated with a higher risk of clinically urgent neurological pathology (CUNP).

MATERIALS AND METHODS

Study setting and participants

This retrospective, multicentre cohort study was carried out in the PED of 11 Italian hospitals (Turin,

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Table 1 Main demographic features and general information of the total cohort and the two subgroups

	Total cohort (n=509)	No CUNP (n=335)	CUNP (n=174)	P value
Age (years)				
Mean (median)±SD	5.8 (4.4)±4.0	5.9±4.1	5.6±3.9	NS
Time before symptom onset (days)				
Mean (median)±SD	3.8 (1.0)±5.4	2.4±3.4	6.4±7.2	<0.001
Sex (%)				
Male	53.6	52.8	55.2	NS
Female	46.4	47.2	44.8	
Triage code (%)				
Red	0.8	0.0	2.3	NS
Yellow	56.0	56.7	54.6	
Green	42.4	42.4	42.5	
White	0.8	0.9	0.6	
Hospitalisation (%)				
Discharge from ED	12.8	19.4	0.0	
Hospitalisation	85.2	77.3	100.0	
Hospitalisation refusal	2.0	3.0	0.0	
Length of stay (days)				
Mean (median)±SD	11.3 (9)±12.20	7.7±5	16.8±17	<0.001
Evolution of symptoms during follow-up (%)				
No information	27.1	31.9	17.8	<0.001
Improvement	63.9	67.5	56.9	
Stability	4.5	0.6	12.1	
Worsening	3.3	0.0	9.8	
Exitus	1.2	0.0	3.4	

The following triage codes were used: red (highly critical conditions), yellow (very urgent), green (urgent) and white (non-urgent).
P values are based on χ^2 and t-test results.
CUNP, clinically urgent neurological pathology; ED, emergency department.

Table 2 Neurological examination findings and reported symptoms in the total cohort and in the two subgroups

	Total cohort (%) (n=509)	No CUNP (%) (n=335)	CUNP (%) (n=174)	P value
Other cerebellar signs*	39.7	37.3	44.3	NS
Positive Romberg sign	37.3	41.2	29.9	0.012
Focal neurological signs	17.5	7.2	37.4	<0.001
Nystagmus	16.5	14.6	0.1	NS
Consciousness impairment	16.1	13.4	21.3	0.023
Hyporeflexia	15.7	8.7	29.3	<0.001
Ophthalmoplegia†	12.0	5.1	25.3	<0.001
Movement disorder	2.9	2.1	4.6	NS
Meningeal signs	2.6	0.6	6.3	<0.001
Papilloedema	2.4	0.0	6.9	<0.001
Seizures	2.0	0.6	4.6	0.004
≥1 associated symptom	80.7	79.7	82.8	NS
Nausea/Vomiting	38.5	37.9	39.7	NS
Headache	29.7	28.1	32.8	NS
Vertigo	24.2	29.0	14.9	<0.001
Fever	23.0	23.9	21.3	NS
Torticollis	2.4	0.9	5.2	0.003
Varicella	16.9	24.8	1.7	<0.001

P values are based on χ^2 test results.
*Dysarthria, tremor or dysmetria.
†Both internal and external.
CUNP, clinically urgent neurological pathology.

Pavia, Padua, Genoa, Bologna, Florence, Siena, L'Aquila, Catania and two centres in Rome). A retrospective medical chart analysis of all patients aged between 12 months and 18 years presenting to the participating PEDs in an 8-year period (January 2009–December 2016) was performed. We included patients with a history of impaired balance and gait incoordination with less than 30 days in duration in whom a clinical diagnosis of ataxia was made in the PED. Ataxia was the predominant sign, or it was clearly the first sign noted at the onset of symptoms. Patients already diagnosed with neurological disorders causing AA were excluded. Gait disorders due to pain, limb weakness, traumatic brain injury or epileptic seizures and patients with severely impaired consciousness were excluded.

Data collection

From each medical record, data on demographic features, clinical history, neurological examination findings, relevant investigations performed (both in the PED and during hospitalisation), hospital admission and length of stay (where applicable) were extracted. The aetiological diagnosis made at the end of the diagnostic work-up was used to classify the cause of AA .

Definitions and outcome measures

Some debate exists over the nosography of parainfectious or postinfectious cerebellar ataxias.[9] In this study, we classified as acute postinfectious cerebellar ataxia (APCA) all cases of postinfectious and parainfectious cerebellar dysfunction. Patients with transitory ataxia without history, signs, symptoms or laboratory

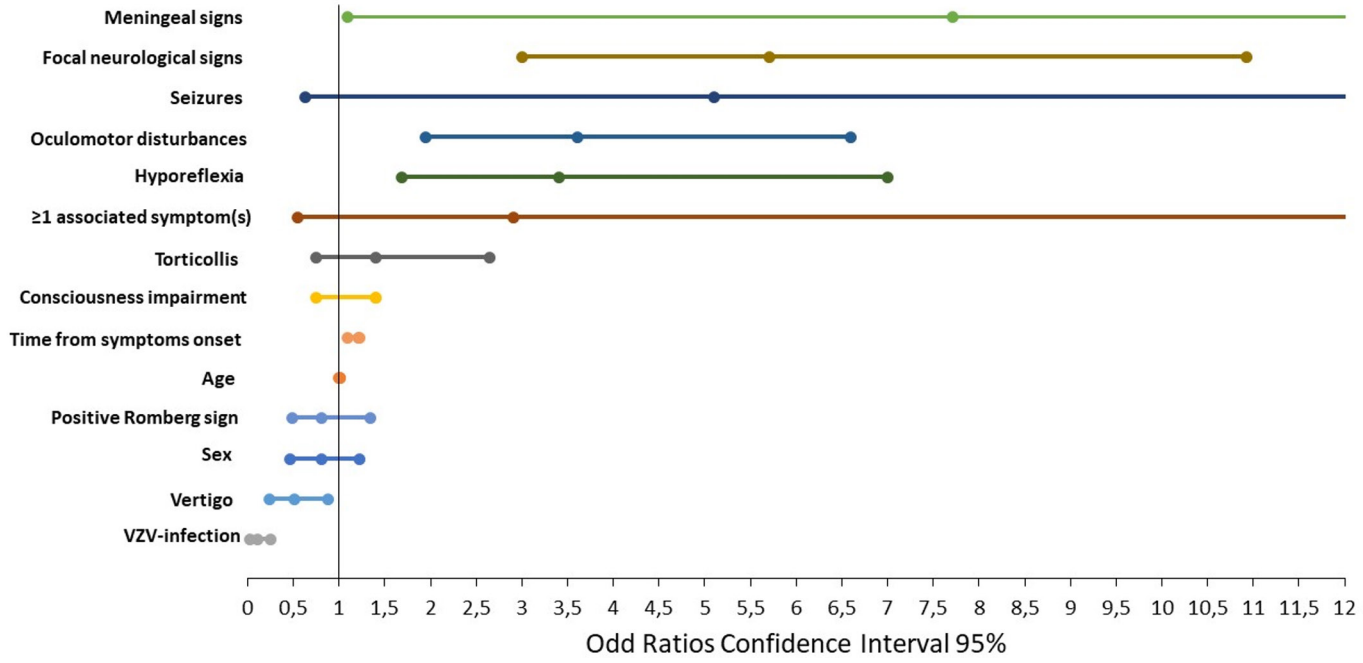

Figure 1 Multivariable regression analysis of predictors of clinically urgent neurological pathology in childhood acute ataxia. 95% CIs for each variable included in the logistic model are shown. Inclusion criteria of candidate clinical predictors are explained in the text. VZV, varicella-zoster virus.

findings of infectious illness were classified as transient undiagnosed ataxia (TUA). Considering that encephalitis, both infectious and autoimmune, may also present with an acute-onset cerebellar syndrome, this term has been reserved to patients developing severely impaired consciousness during the illness or showing widespread cerebral involvement on neuroimaging, diffuse electroencephalographic slowing or (in case of infectious encephalitis) microbiological evidence of direct infection of the central nervous system (CNS).

Psychogenic ataxia was diagnosed based on the presence of a phenomenology incongruent with an organic disorder, evidence of suggestibility, distractibility and/or variability of symptoms or inconsistent disability.

For the purposes of our study, CUNP was defined as any nervous system disorder requiring early diagnosis and prompt medical or surgical interventions to prevent disabling or life-threatening evolution, namely neoplastic, cerebrovascular and infectious CNS disorders, demyelinating diseases, acute neuropathies (AN), genetic or metabolic disorders, and CNS malformations requiring surgical treatment.

Statistical analysis

We described the clinical and demographic features of the overall cohort and of the two diagnostic subgroups (patients with and without CUNP). Each variable was tested to identify significant differences between the two subgroups. After reviewing for appropriateness, χ^2 and Student's t-tests were used for categorical and continuous variables, respectively.

We applied a logistic regression analysis model to assess the presence of any predictive variables associated with a higher risk of CUNP. Inclusion of variables in the model was based on clinical plausibility and significant or nearly significant differences on χ^2 and t-tests. Variables with extremely unbalanced distribution in the two groups (frequency of 0% in one group) were excluded. Adjusted OR and 95% CI were used as measures of effect. Statistical significance was set at $p<0.05$.

In addition, we performed a multivariable regression analysis to evaluate the clinical factors responsible for an increased physicians' attitude to request neuroimaging. Each clinical feature was tested for significant differences between patients who performed and did not perform neuroimaging. Mann-Whitney, χ^2 and Student's t-tests were used, where appropriate. All variables showing significant differences, together with sex and age, were included in the model.

Finally, to evaluate indication to perform neuroimaging, cerebrospinal fluid (CSF) sampling and nerve conduction study (NCS), selected clinical features were compared in patients with normal and abnormal findings. Mann-Whitney, χ^2 and Student's t-tests were used, where appropriate.

IBM SPSS Statistics V.24.0 software was used to perform all statistical analyses.

RESULTS

Main clinical and demographic features

During the study period, 2 426 030 children were admitted to the participating PEDs. A total of 509 patients with AA (male:female=1.16) were included, with a mean age of 5.81 years. The frequency of AA was of 1 case for every 4766 emergency department attendances (0.021%). The main demographic features and clinical findings of the total cohort and the two subgroups are summarised in tables 1 and 2, respectively. Investigations performed are illustrated in online supplementary table S1.

Associated symptoms and neurological exam findings

On admission, 80% of the patients reported some associated symptoms, mainly nausea or vomiting (38.5%) and vertigo (24.2%). Twenty-three per cent of the children reported febrile illness or were febrile on admission (table 2). On neurological examination, additional cerebellar signs were the most frequently reported neurological abnormalities (39.7%), followed by positive Romberg sign (37.3%).

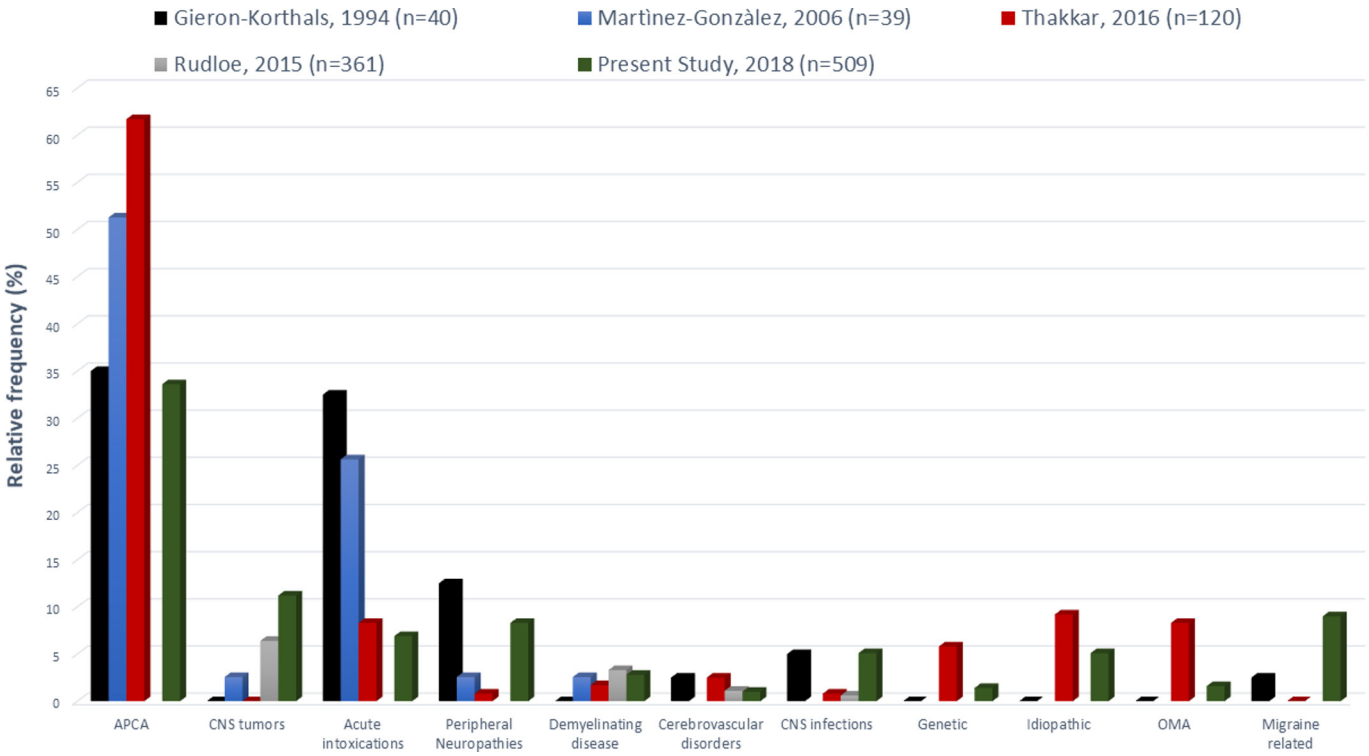

Figure 2 Comparative representation of distribution of the main causes of acute ataxia in the different cohorts reported in the literature. Due to the study design, only the frequency of disorders causing significant neuroimaging abnormalities was deducible from the study by Rudloe *et al*[6]. APCA, acute postinfectious cerebellar ataxia; CNS, central nervous system; OMA, opsoclonus-myoclonus-ataxia.

Ataxia causes

The most commonly identified cause of AA was APCA (33.6%) (online supplementary figure S2A), mainly due to varicella-zoster virus (VZV) infection (49.7%; online supplementary figure S2B,C). In the remaining cases, the triggering infection was isolated only in a minority of patients (29.1% of non-VZV-related APCA; online supplementary figure S2B). The second most common cause of AA was CNS tumours (57 cases, 11.2%).

Acute encephalitis was diagnosed in 30 children (5.9%), mostly infectious (23 cases), followed by autoimmune and paraneoplastic forms. ANs were found in 42 patients (8.3%).

Migraine-related disorders such as vestibular migraine and benign paroxysmal vertigo (BPV) were diagnosed in 46 patients (27 and 19 cases, respectively), representing the third most common group (9%). Psychogenic ataxia was recognised in 21 cases (4.1%), and 35 cases of acute intoxications due to drug overdose or substance abuse were found (online supplementary figure S4). Other less frequently encountered causes were vestibular disorders (5.3%), demyelinating diseases (2.8%), genetic-metabolic disorders (1.4%) and CNS malformations (0.8%; online supplementary figure S2A). Twenty-six patients (5.1%) were classified as TUA.

Logistic regression model

Based on the final diagnosis, 174 patients with CUNP were identified. Following comparison between patients with and without CUNP (tables 1–2), 14 variables were included in the model (figure 1, online supplementary table S2).

Papilloedema, the only finding appearing exclusively in CUNP patients, was excluded from the logistic analysis. Meningeal and focal neurological signs were associated with a higher risk of CUNP adjusting for other variables, followed by hyporeflexia and ophthalmoplegia. The odds of an underlying CUNP were increased by 51% by each day from onset of ataxia. Conversely, a history of VZV infection and vertigo resulted in a lower risk of CUNP. Adjusting for other variables, neither seizures nor consciousness impairment was significantly associated with CUNP.

Diagnostic investigations

Neuroimaging was performed in 351 children (69%), with abnormal findings in 129. Neuroimaging was more frequently requested in patients with other cerebellar signs, oculomotor deficits, nystagmus, focal signs, papilloedema and longer duration of symptoms (online supplementary table S3A), and less frequently performed in patients with history of VZV infection. Even when adjusting for other variables, the stronger predictors of neuroimaging use were the presence of other cerebellar signs, ophthalmoplegia and focal neurological signs. In addition to a history of varicella, longer duration of evolution of symptoms was also associated with a lower probability of being investigated with neuroimaging (online supplementary table S3B).

Among patients undergoing neuroimaging, abnormal results were significantly more frequent in patients with focal neurological signs, torticollis, consciousness impairment, papilloedema and ophthalmoplegia ($p<0.03$; online supplementary table S4). Conversely, they were less frequent in patients with varicella, vertigo or fever ($p<0.02$). Patients with abnormal neuroimaging showed longer time from symptom onset ($p<0.001$).

NCS was performed in 42 children; alterations were more common in children with hyporeflexia ($p<0.001$; online supplementary table S5). CSF samples, collected in 109 patients (21.4%), were significantly more frequently altered in children with hyporeflexia ($p=0.003$) and in those with focal neurological signs ($p=0.015$; online supplementary table S6). Conversely,

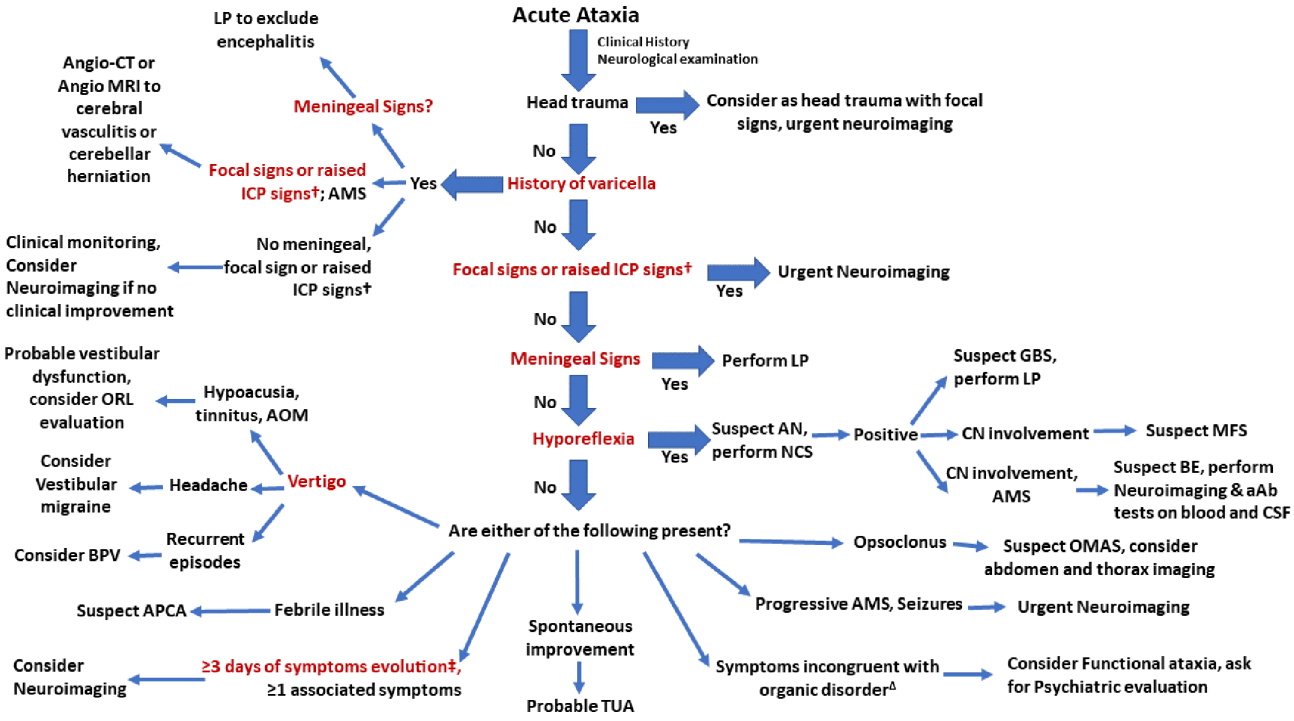

Figure 3 Flow-chart for the diagnostic evaluation of the acutely ataxic child. Signs and symptoms significantly associated with a higher risk of CUNP in our model are marked in red. †Raised ICP signs: papilledema, ophthalmoplegia, mydriasis, vomiting without nausea. ‡The cut-off of 3 days is indicated basing on the risk stratification model proposed by Rudloe *et al*.[6] ΔEvidence of suggestibility, distractibility and/or variability of symptoms or inconsistent disability. aAB, autoantibodies; AMS, altered mental status; AN, acute neuropathy; AOM, acute otitis media; APCA, acute post-infectious cerebellar ataxia; BE, Bickerstaff encephalitis; CSF, cerebrospinal fluid; CN, cranial nerve; GBS, Guillain Barré Syndrome; ICP, intracranial pressure; LP, lumbar puncture; MFS, Miller Fisher syndrome; NCS, nerve conduction study; OMAS, opsoclonus-myoclonus-ataxia syndrome; ORL, otorhinolaryngology; TUA, transient undiagnosed ataxia.

the presence of other cerebellar signs was associated with a lower frequency of CSF alterations (p=0.002).

DISCUSSION

AA epidemiology

Few data exist about the real incidence and prevalence of AA in childhood.[4] Our study represents the first, large multicentre study on AA conducted in a paediatric emergency setting, showing a frequency of 0.02% of all emergency department attendances, similar to previous PED-based studies.[5 6] Figure 2 shows the differences in frequency of the main causes of AA between our cohort and those previously reported.

APCA was the most frequent aetiology encountered. As previously described in the Italian milieu,[10] VZV is the predominantly involved infectious agent, probably because of the still poor, fluctuating and heterogeneous vaccination coverage all over the country (online supplementary figure S3[11]). Given that the spread of VZV vaccination is expected to diminish the incidence of VZV complications (including APCA[12]), the diagnostic approach to AA should be re-evaluated in light of an evolving immunisation context, as already suggested in the USA.[6]

Probably, the inclusion of children with up to 30 days of symptom evolution explains the relatively high frequency of CNS tumours and the low frequency of acute intoxications, compared with previous reports[5–8] (figure 2). Of note, almost one-third of acute intoxications were due to substance abuse, suggesting that it should always be suspected in otherwise unexplained AA, even in a paediatric setting.

In our cohort, we noted a high occurrence of vestibular migraine and BPV, almost absent in the previous studies.[5–8] The relationship between these two entities is yet unclear,[13] but they may be an increasingly recognised cause of AA in the future. Also AN was frequently encountered in our series (8.3%), with 36 cases of Guillain-Barré syndrome, suggesting that AA is a common presentation of this condition in children.

Noteworthy, a definite aetiology was not identified in a considerable proportion of patients with transient symptoms. The frequency of undiagnosed cases was highly variable in the previous studies, ranging from 9.2% to apparently no cases[5 7 8] The lack of undiagnosed cases in the smaller cohorts is largely imputable to chance. In addition, other factors probably play a role, such as the study setting (the higher the care level, the higher the proportion of undiagnosed patients due to the selection of rare causes) or the criteria applied for clinical diagnosis of several entities, namely APCA or psychogenic disorders (the stricter the criteria, the larger the number of unexplained cases).

Risk factors for urgent conditions

The second aim of our study was to determine the clinical features predictive of an underlying pathology requiring early intervention. Given its self-limiting evolution and good outcome, APCA was not considered as a CUNP. According to our model, children with a recent history of varicella or vertigo can be allocated to a low-risk category, these two features, respectively, pointing to VZV-related APCA and vestibular dysfunction. As expected, the presence of focal or meningeal signs, hyporeflexia and ophthalmoplegia is strongly suggestive of CUNP. A longer duration of symptoms before PED evaluation emerged as a significant risk factor for CUNP, accordingly

with the risk stratification model proposed by Rudloe *et al*[6] for identifying patients with intracranial pathology. This is probably explained by the insidious onset of some severe ataxia causes, such as brain tumours, showing how the emergency health service often intercepts patients with subacute and severe pathologies that require a high level of suspicion to avoid misdiagnosis. As cited above, papilloedema, a clear indication for neuroimaging, was not included in our model. Interestingly, extremely concerning findings such as consciousness impairment and seizures, despite their positive association with CUNP, did not reach significance. Several reasons can explain this finding. First, CUNP (although it is of interest for emergency physicians) is a broad and heterogeneous category, only defined by the urgent characteristics. Many urgent conditions presenting with ataxia do not cause consciousness impairment (eg, AN, posterior fossa tumours), and many non-urgent conditions (eg, migraine or functional disorders) may present with (or may mimic) an altered mental status. This weakens the association between consciousness impairment and CUNP. With regard to seizures, they are an infrequent event in patients with ataxia, its frequency probably being insufficient to reach significance in the multiple regression analysis. With the aim of a real-life application of our findings, we suggest that any child with a symptom or sign with an OR >1 should be appropriately investigated. On this basis, we developed a flow chart of a diagnostic approach to evaluating a child with AA (figure 3).

Diagnostic investigations in a child with ataxia

As expected, neuroimaging alterations are significantly more frequently found in patients with signs of CNS involvement (focal neurological signs, consciousness impairment, papilloedema, ophthalmoplegia, head tilt), but also in patients with longer evolution of symptoms. Noteworthy, most patients without CUNP underwent neuroimaging studies, configuring an overuse of diagnostic testing possibly exposing patients to unnecessary risks.[14] Hyporeflexia appears to be the most suggestive finding of AN.

Study limitations

The present study suffers from some limitations, mostly related to its retrospective nature. First, the accuracy of data is dependent on the physician's experience. In our study, neurological evaluation was performed by emergency physicians, who are not expected to be experienced in neurological assessment. Although neurologist consultation was requested in many cases (65.6%), the robustness of clinical assessment could have been partially limited. In addition, some clinical information may have not been reported correctly on emergency department records. Other limitations are due to exposure to some selection biases. In fact, some AA cases could have been misdiagnosed, underestimating their prevalence. By contrast, given the tertiary nature of most participating centres, the frequency of AA could have been overestimated at this care level. Finally, the CUNP category has been designed aiming to help clinicians in identifying patients with urgent needs, which is a primary concern in an emergency setting. However, it is not informative about the risk of a specific underlying condition.

CONCLUSIONS

Our study demonstrates that AA is an infrequent but concerning neurological emergency in childhood. The most frequent cause is APCA, but CUNPs account for over a third of the AA cases encountered in the PED. Focal and meningeal signs, hyporeflexia and ophthalmoplegia, as well as longer duration of symptoms, are the most consistent 'red flags' of a severe underlying pathology. Other features with less robust association with CUNP, such as seizures or consciousness impairment, should be seriously taken into account during AA evaluation.

Author affiliations
[1]University Hospital Pediatric Department, Bambino Gesù Children's Hospital IRCCS, University of Rome Tor Vergata, Rome, Italy
[2]Pediatric Emergency Department, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy
[3]National Centre for Epidemiology, Surveillance and Health Promotion, National Institute of Health, Rome, Italy
[4]Chair of Pediatrics, NESMOS Department, Faculty of Medicine and Psychology, Sapienza University of Rome, Sant'Andrea Hospital, Rome, Italy
[5]Department of Pediatric Emergency, Regina Margherita Pediatric Hospital, Turin, Italy
[6]Child Neurology and Clinical Neurophysiology, Department of Woman and Child Health, University of Padua, Padova, Italy
[7]Department of Medical Sciences, Pediatric Section, University of Ferrara, Ferrara, Italy
[8]Pediatric Emergency Department, Giannina Gaslini Children's Hospital, Genoa, Italy
[9]Pediatric Emergency Unit, Anna Meyer Children's Hospital, Florence, Italy
[10]Child Neurology Unit, University of Bologna, Bologna, Italy
[11]Department of Pediatrics, Fondazione Policlinico San Matteo IRCCS, University of Pavia, Pavia, Italy
[12]Clinical Pediatrics, Department of Molecular Medicine and Development, University of Siena, Siena, Italy
[13]Pediatric Emergency Unit, Department of Pediatrics, Child Neurology and Psychiatry, Sapienza University of Rome, Rome, Italy
[14]Unit of Pediatrics and Emergency Pediatrics, AOU Policlinico Vittorio Emanuele, Catania, Italy
[15]Department of Pediatrics, University of L'Aquila, L'Aquila, Italy
[16]Department of Pediatrics, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy
[17]Pediatric Emergency Department, Department of Woman's and Child Health, University of Padova, Padova, Italy
[18]Division of Neurology, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy

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Contributors UR and GG conceptualised and designed the study, coordinated and supervised the data collection, interpreted the data, drafted the initial manuscript, provided critical review and revision of the manuscript, and wrote the final manuscript. NV performed the statistical analysis, interpreted the data, contributed to conceptualising the study and participated in the design of the study, and reviewed and revised the initial manuscript. ARe and PP contributed to conceptualising the study and participated in the design of the study, interpreted the data, and reviewed and revised the initial manuscript. CB, AS, GB, LC, DMC, SS, SG, FM, RF, AV, EB, CV, LDD, RM, SMas, LM, TF, AR, CG, SMar, CDP, LP, AFU and RR contributed to conceptualising the study, collected the data, and reviewed and revised the initial manuscript. All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

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Patient consent for publication Not required.

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