RESEARCH

Pediatric Dystonic Storm

A Hospital-Based Study

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Abstract

Objective

Pediatric dystonic storm is an underrecognized entity. We aimed to evaluate the profiles of children presenting with dystonic storm in a referral hospital. Management schema and treatment responsiveness of this uncommonly reported entity were analyzed.

Methods

Retrospective review of all children (up to 18 years) hospitalized with dystonic storm over 39 months in the aforementioned facility.

Results

Twenty-three children whose ages ranged from 2 years 2 months to 14 years 4 months years (median: 6 years 11 months) (males: 13, females: 11) presented with dystonic storm. The annual incidence was 0.4 per 1,000 fresh admissions with an event rate of 0.9 per 1,000 for all admissions. All had Dystonia Severity Action Plan grades 4/5 with identifiable trigger in 13 (50%). Underlying dystonic disorder preexisted in 10 (43.4%); 8 of these had cerebral palsy. Polypharmacotherapy with >4 drugs out of trihexyphenidyl, tetrabenazine, clonazepam, gabapentin, levodopa-carbidopa, trichlorophos, and melatonin was needed. Supportive care and adequate sedation helped in symptom control. All children were managed with midazolam infusion over 2–10 days (median: 5 days). Mechanical ventilation was resorted to in 6 children (3–22 days). Vecuronium and propofol were used in 3/23 (13%) and 4/23 (17%) children, respectively. Deep brain stimulation was curative in 1 child. Hospitalization ranged from 5 to 31 (median: 11) days. Although there were no deaths, rhabdomyolysis was noted in 1 child. Postdischarge, 6 (26%) children relapsed.

Conclusions

Dystonic storm is a medical emergency mandating aggressive multimodal management. Supportive care, antidystonic drugs, and early elective ventilation alongside adequate sedation with benzodiazepines ameliorate complications. Relapses of dystonic storm are not uncommon.

Dystonic storm, also known as status dystonicus and dystonic crisis, is an emergency movement disorder.[1] It remains underdiagnosed and underreported.[2,3] It remains a highly

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distressing condition for parents of affected children in the absence of an effective and timely treatment. Management of dystonic storm demands multimodal approach in an intensive care setup with close monitoring for complications.[4] No consensus guideline for management of this medical emergency exists. The present study seeks to estimate the burden of this entity among pediatric hospitalizations and analyze the clinical profile, management schema, and response variables of this vexing entity.

Methods

This study occurred in a quaternary care level armed forces medical services superspecialty hospital, which forms the final referral node for a network of medical facilities spread all over India. The hospital information system was searched for a diagnosis of dystonia/dystonic disorder/dystonic storm/status dystonicus from January 2017 till March 2020 with a filter of below 18 years of age. Medical records of all these children were retrieved and analyzed. Any child younger than 18 years admitted with an episode of dystonic storm was included in the study. Dystonic storm was defined as a severe, hyperkinetic movement disorder, which is life threatening and mandates emergency management.[1,4] Clinical features were tabulated along with management strategies and treatment responsiveness. Data capture and statistical analysis were performed using Microsoft Excel.

Standard Protocol Approvals, Registrations, and Patient Consents

a) Approval by an ethical standards committee on human experimentation (institutional or regional) for any experiments using human participants was not obtained, as the study was a retrospective one and did not involve human experimentation in any manner.
b) Institutional ethical committee approval was obtained for the study.
c) Authorization has been obtained for disclosure (consent to disclose) of any recognizable persons in any information that may be published in the Journal, in derivative works by the AAN, or on the Journal's website.

Data Availability

Individual deidentified participant data pertaining to the demographic variables, baseline drugs, therapies administered, and outcomes shall be shared.

Results

During the 39-month study period, 33 episodes of dystonic storm were identified among the 11,336 total pediatric admissions. These occurred among 23 children among the 16,632 children reporting to pediatric neurology out-patient department during this period and subsequently getting hospitalized. This leads to an annual incident rate of 0.4 per 1,000 fresh admissions below 18 years age and event rate of 0.9 per 1,000 per year for all admissions including recurrent hospitalizations. The demographic profile of the cases is presented in table 1. The ages of children ranged between 2 years 2 months to 14 years 4 months (median: 6 years 11 months). No significant difference in numbers of known dystonics on pediatric neurology follow-up vs those presenting de novo with dystonic storm was noted (n = 10 vs n = 13) ($p = 0.38$). There was no sex predilection (males: 13; females: 11; $p = 0.56$). The majority (19%) of children had Dystonia Severity Action Plan (DSAP) grade 4; 4 had DSAP grade 5 at admission. Children were worked up for etiology during their initial presentation as per prevailing standard guidelines. Neuroimaging was performed in all, whereas other investigations such as metabolic screening and genetic testing were ordered only if indicated. When children on follow-up were admitted with dystonic crisis, fresh investigations were ordered keeping in view possible impending metabolic derangements. Repeat neuroimaging was not performed.

About a third of these children had cerebral palsy (CP), whereas the rest had a multitude of conditions. Only 1 child had a primary genetically proven dystonia. The underlying condition in 1 child (serial no. 11, table 1) was not established despite extensive investigations.

Dystonic storm occurred in 10/23 children (43%) while on antidystonic drug therapy (table 2, figure 1). A precipitating trigger was identifiable in 10/23 (43%) children (table 1). Precipitants included infectious trigger in 8 (fever without focus: 4; upper respiratory tract infection: 1; diarrhea: 1; encephalitis: 2), whereas 2 children had drug-related trigger (poor compliance: 1; change in drug brand: 1) (table 1). Most children (21/23; 91%) had generalized dystonia irrespective of underlying etiology. Predominant oromandibular and limb dystonia were noted in 2 children at presentation. Both of them had autoimmune encephalitis (anti-NMDA receptor antibody encephalitis: 1; seronegative autoimmune encephalitis: 1) (patient nos. 2 and 17, table 1). Mixed dystonia was the commonest, whereas predominant tonic dystonia and predominant phasic dystonia occurred in 5 and 6 children, respectively. Both children with autoimmune encephalitis had phasic dystonia. Concomitant cognitive regression with phasic dystonia along with EEG and CSF antimeasles antibody titers led to a diagnosis of subacute sclerosing panencephalitis (SSPE) in 1 case. All 3 children with underlying neurometabolic disorder (pantothenate kinase deficiency, Wilson disease, and glutaric aciduria type 1) had pronounced phasic dystonia.

Management included symptomatic pharmacotherapy and supportive care with adequate hydration, nutrition, and respiratory support (tables 1 and 2). Mechanical ventilation was resorted to in 6/23 (26%) children for variable duration ranging 3–22 days (figure 2). No ventilator-related complication was recorded. Rhabdomyolysis was documented in a male child with Dravet syndrome on day 3 of admission.

Table 1 Demographic Profile of the Study Population

Patient serial number	Age (y)	Sex	Diagnosis	Trigger	Timing of presentation	Baseline drugs	Predominant type of dystonia	Parts of body affected	Complication	Total hospitalization (d)
1	8	Female	AADC deficiency	Fever without focus	I	No	Phasic	Generalized	No	5
2	6.1	Female	Autoimmune encephalitis	None	I	No	Phasic	Oromandibular + limb	No	7
3	2.2	Male	Dystonic CP	Respiratory tract infection	FU	Trihexyphenidyl	Tonic	Generalized	No	7
4	3	Female	Mixed CP	Poor drug compliance	FU	Trihexyphenidyl, clonazepam, and baclofen	Tonic	Generalized	No	7
5	3.2	Female	Mixed CP	None	FU	Trihexyphenidyl, clonazepam, and baclofen	Phasic	Generalized	No	8
6	4.2	Male	Dystonic CP	None	FU	Trihexyphenidyl	Tonic	Generalized	No	8
7	5.8	Male	Dyskinetic CP	Drug brand change	FU	trihexyphenidyl, gabapentin, and clonazepam	Mixed	Generalized	No	8
8	6.6	Male	Dystonic CP	Fever without focus	FU	Trihexyphenidyl	Mixed	Generalized	No	9
9	10.5	Female	Mixed CP	None	FU	Trihexyphenidyl, clonazepam, and baclofen	Mixed	Generalized	No	9
10	14.4	Female	Dystonic CP	None	FU	Trihexyphenidyl	Mixed	Generalized	No	9
11	4.7	Male	Idiopathic	None	I	No	Mixed	Generalized	No	10
12	3.8	Male	Dravet syndrome	Fever without focus	I	Valproate and clobazam	Tonic	Generalized	Yes (rhabdomyolysis)	11
13	12.6	Male	Herpes simplex virus encephalitis	Encephalitis	I	No	Mixed	Generalized	No	11
14	6.11	Female	Glutaric aciduria type 1	Diarrhea	I	No	Phasic	Generalized	No	11
15	11.2	Female	Idiopathic	None	I	No	Phasic	Generalized	No	11
16	9	Male	Japanese encephalitis	Encephalitis	I	No	Phasic	Generalized	No	12
17	4.4	Female	Anti-NMDAR antibody encephalitis	None	I	No	Phasic	Oromandibular + limb	No	13
18	8	Male	PKAN	None	I	No	Phasic	Generalized	No	13
19	6.4	Female	Postencephalitic sequelae	Fever without focus	I	No	Phasic	Generalized	No	15
20	9.9	Female	Head trauma sequelae	None	I	No	Tonic	Generalized	No	18
21	4.11	Male	Primary dystonia	None	FU	Tetrabenazine and clobazam	Phasic	Generalized	No	23
22	2.3	Male	SSPE	None	I	No	Phasic	Generalized	No	26
23	7.1	Male	Wilson disease	None	FU	D-penicillamine and zinc	Phasic	Generalized	No	31

Abbreviations: AADC = aromatic L-amino acid decarboxylase; CP = cerebral palsy; FU = on follow-up; I = initial presentation; NMDAR = NMDA receptor; PKAN = pantothenate kinase-associated neurodegeneration; SSPE = subacute sclerosing panencephalitis.

Table 2 Management Details of the Study Population

Patient serial number	Duration of midazolam infusion (d)	Midazolam infusion rate (max) (μg/kg/min)	Duration of vecuronium infusion (d)	Duration of propofol infusion (d)	Trihexyphenidyl (n:23)	Clonidine (n:21)	Tetrabenazine (n:17)	Melatonin (n:23)	Trichlorophos (n:23)	Gabapentin (n:23)	Rhabdomyolysis (n:1)	Additional drugs (n:6)	Adverse effect of drugs (n:5)	DBS (n:1)
1	2	7	0	0	Yes	Yes	Yes	Yes	Yes	Yes	No	Nil	Hypotension (midazolam dose reduced)	No
2	2	3	0	0	Yes	Yes	Yes	Yes	Yes	Yes	No	IV immunoglobulin and methylprednisolone	Hypotension	No
3	3	30	2	1	Yes	Yes	Yes	Yes	Yes	Yes	No	Nil	Hypotension	Yes
4	3	20	0	2	Yes	Yes	Yes	Yes	Yes	Yes	No	Nil	Nil	No
5	4	5	0	0	Yes	Yes	Yes	Yes	Yes	Yes	No	Nil	Nil	No
6	4	3	0	0	Yes	Yes	Yes	Yes	Yes	Yes	No	Nil	Nil	No
7	4	5	0	0	Yes	Yes	No	Yes	Yes	Yes	No	Nil	Nil	No
8	4	6	0	0	Yes	Yes	No	Yes	Yes	Yes	No	Nil	Nil	No
9	5	5	0	0	Yes	Yes	Yes	Yes	Yes	Yes	No	Nil	Nil	No
10	5	3	0	0	Yes	Yes	Yes	Yes	Yes	Yes	No	Nil	Nil	No
11	5	5	0	0	Yes	Yes	No	Yes	Yes	Yes	No	Nil	Nil	No
12	5	18	3	0	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Valproate	Nil	No
13	6	4	0	0	Yes	Yes	Yes	Yes	Yes	Yes	No	Acyclovir	Nil	No
14	6	2	0	0	Yes	Yes	Yes	Yes	Yes	Yes	No	Carnitine, coenzyme Q, thiamine, and riboflavin	Nil	No
15	6	15	0	0	Yes	Yes	Yes	Yes	Yes	Yes	No	Nil	Nil	Yes
16	6	7	0	2	Yes	Yes	No	Yes	Yes	Yes	No	Nil	Nil	No
17	7	22	5	1	Yes	No	Yes	Yes	Yes	Yes	No	IV immunoglobulin and methylprednisolone	Hypotension (midazolam reduced, dopamine administered)	No
18	7	3	0	0	Yes	No	Yes	Yes	Yes	Yes	No	Nil	Nil	No
19	7	5	1	2	Yes	Yes	Yes	Yes	Yes	Yes	No	Nil	Nil	No
20	9	3	0	0	Yes	Yes	Yes	Yes	Yes	Yes	No	Nil	Nil	No
21	9	7	0	0	Yes	Yes	No	Yes	Yes	Yes	No	Valproate	Nil	No

Continued

Table 2 Management Details of the Study Population *(continued)*

Patient serial number	Duration of midazolam infusion (d)	Midazolam infusion rate (max) (μg/kg/min)	Duration of vecuronium infusion (d)	Duration of propofol infusion (d)	Trihexyphenidyl (n:23)	Clonidine (n:21)	Tetrabenazine (n:17)	Melatonin (n:23)	Trichlorophos (n:23)	Gabapentin (n:23)	Rhabdomyolysis (n:1)	Additional drugs (n:6)	Adverse effect of drugs (n:5)	DBS (n:1)
22	9	18	0	0	Yes	Yes	No	Yes	Yes	Yes	No	Carbamazepine and isoprinosine	Hypotension (midazolam dose reduced)	No
23	10	3	0	0	Yes	Yes	Yes	Yes	Yes	Yes	No	D-penicillamine and zinc	Nil	No

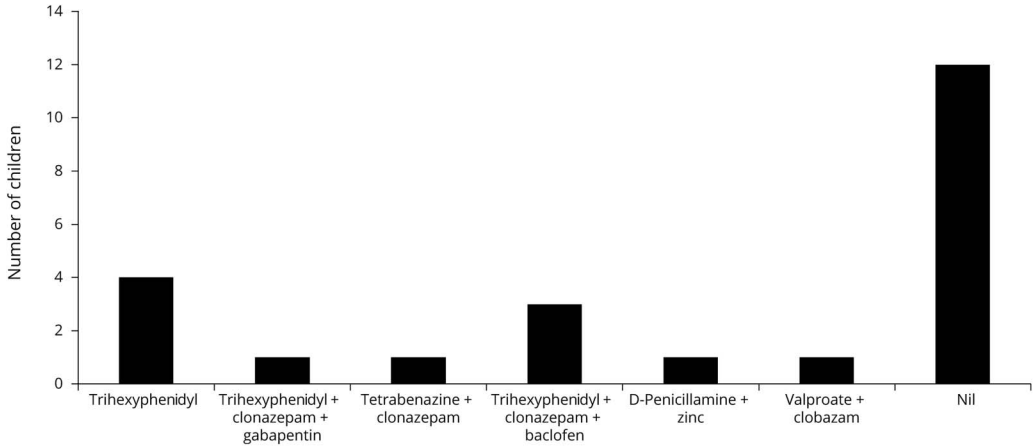
Abbreviation: DBS = deep brain stimulation.

Child was managed with adequate hydration, and he made an uneventful recovery by 24 hours.

Midazolam infusion was the cornerstone of management. A standardized institutional protocol was followed consisting of an early initiation of continuous midazolam infusion at admission titrated to achieve cessation of dystonia followed by tapering on achieving DSAP grade 3 and switch to an intermittent dosing schedule and subsequent substitution with oral clonazepam (figure 3). Midazolam infusion was continued for 2–10 days (median [interquartile range] 5 [4–7] days). The peak continuous infusion rate ranged from 2 to 30 μg/kg/min (median [interquartile range]: 5 μg/kg/min [3–14.5]) (table 1, figure 2). Hypotension occurred in 4 children, which necessitated tapering of the infusion in 3 children (13%) and addition of dopamine infusion 1 child. Inj. vecuronium was administered in 4 children (17%) for muscle relaxation. Propofol was used in 3 children (13%), which were tapered off by 36 hours (table 1). All children were simultaneously exhibited multiple antidystonia drugs (table 2). Injectable botulinum toxin was not exhibited to children with focal dystonia (patient nos. 2 and 17; table 1). These children had autoimmune encephalitis wherein evidence of injectable botulinum toxin therapy is sparse.

Fluctuations in dystonia were noted in every child, occasionally mandating increase in dosage of midazolam. Drugs were administered round the clock with adequate response, acceptability, and adverse effects. Two children of 21 (9%) exposed to clonidine had hypotension requiring discontinuation of drug. Exacerbations of dystonia were effectively controlled using intermittent dosing of melatonin/triclophos. This phenomenon was observed in every child even when on midazolam infusion while it was being tapered. Although midazolam was being tapered, children were also exhibited clonazepam/nitrazepam round the clock through oral route/nasogastric tube (figure 3), the dosing of which was guided based on symptom control and drug tolerance. Oral medications that were continued simultaneously included trihexyphenidyl, in certain cases depending on symptom responsiveness. As per the institutional protocol, trihexyphenidyl and gabapentin were exhibited universally, whereas tetrabenazine, gabapentin, clonidine, baclofen, and levodopa-carbidopa were prescribed sequentially based on the individual's symptom refractoriness. Levodopa-carbidopa and tetrabenazine were not administered simultaneously in view of antagonistic modes of action. However, baseline drugs, which a child was receiving before hospitalization, were continued in deference to the aforementioned protocol. Additional therapies were exhibited in children presenting with dystonic crisis due to defined etiologies (2 children with autoimmune encephalitis (IV immunoglobulin and methylprednisolone pulse followed by oral prednisolone), 1 child with Wilson disease (D-penicillamine and zinc), glutaric aciduria type 1 (treated with carnitine, coenzyme Q, and thiamine), herpes simplex encephalitis (IV acyclovir), and SSPE (carbamazepine and isoprinosine). All children were discharged after attaining DSAP grade 1. Duration of hospitalization ranged from 5 to 31 days (median:

Figure 1 Drugs Taken by the Children Before Initial Admission for Dystonic Storm



11 days) (table 2). Children with CP needed a significantly lesser duration of midazolam infusion and had a significantly short period of hospitalization. On follow-up, 6 children had relapse of dystonic storm (table 3). All drug formulations were generic in nature and manufactured in India. The underlying illness in these children was varied. For instance, a child with CP diagnosed on the basis of clinical history and neuroimaging (serial no. 2, table 3) had multiple relapses. Investigations for genetic/metabolic etiologies were omitted in the aforementioned child in view of supportive clinical history and neuroimaging as per prevailing institutional policy. However, another child with relapsing dystonic crisis (serial no. 4, table 3) had undergone an extensive battery of investigations including whole-exome sequencing with no diagnostic yield. No child died during the study period.

Discussion

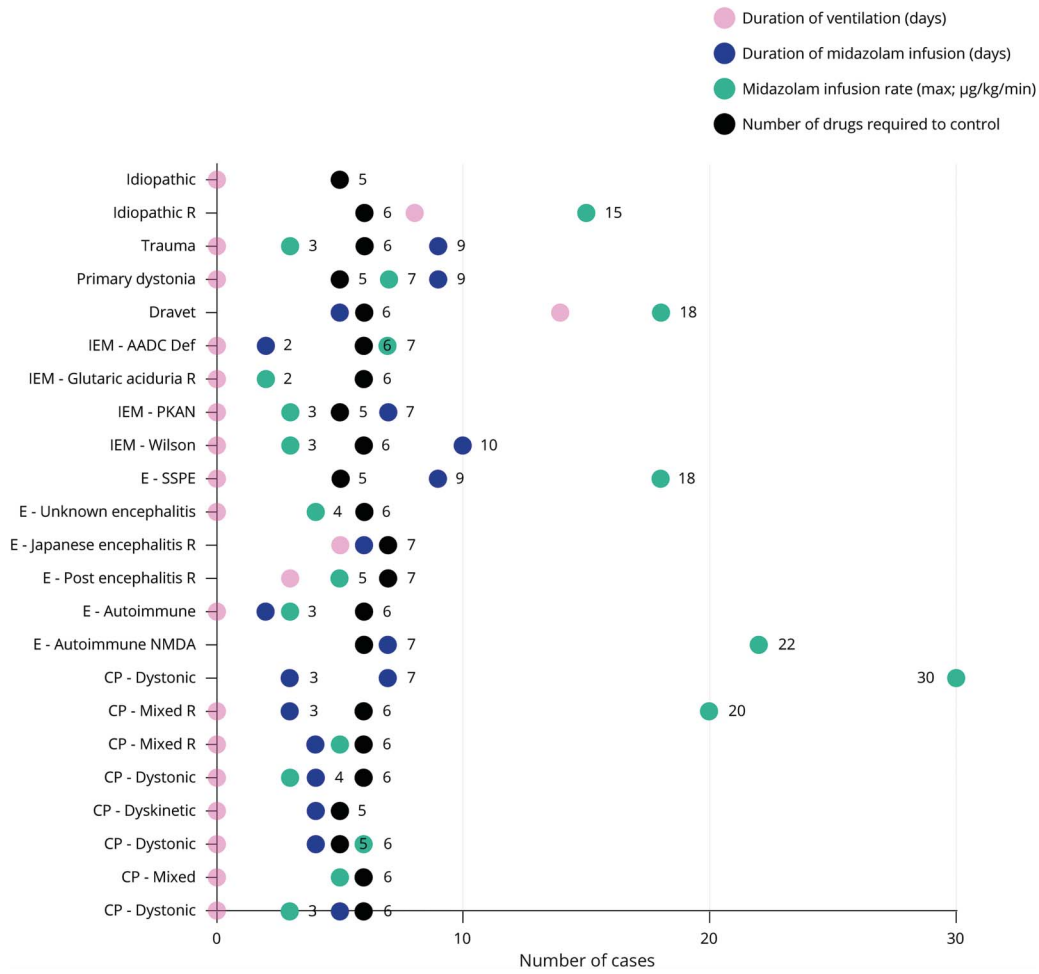
Dystonic storm is infrequently reported in the pediatric literature. There are ~100 case reports among all age groups with few large series.[1] Actual incidence is likely to be higher owing to an underdiagnosis by most clinicians.[1] Our prevalence estimates suggest that it is not an uncommon entity in a pediatric neurology service. Akin to the published literature, no sex predilection at presentation was noted by us.[5]

The number of children who presented initially with dystonic storm was similar to those who were already on follow-up for dystonia and had acute exacerbation. Above observation may possibly be attributed to the underlying etiologies. The majority of these (8/23) on follow-up for dystonia had CP, whereas 1 child each had Wilson disease and primary dystonia. Fasano et al.[5] studied a total of 89 episodes of dystonic storm in 68 patients, wherein 37.8% of children with secondary dystonia had CP. CP comprised 35% of children who were on follow-up for dystonia in our series too.

An initial presentation with dystonic storm should prompt clinicians to actively look for underlying etiologies. Dystonic storm may be the initial presentation at varying ages. Children who presented de novo for the first time had varied etiologies including infective (encephalitis), immune-mediated (autoimmune encephalitis), neurotransmitter disorder, metabolic, and posttraumatic. In 1 child with SSPE with dystonic storm, the phasic dystonia was indicative of an epileptic event and responded to carbamazepine. Children presenting with autoimmune encephalitis and herpes simplex virus encephalitis also responded to specific treatment modalities.

Fever without focus was the commonest trigger in our study. Other common trigger factors previously reported include gastroenteritis, medication adjustment, and constipation.[1,6] About half of children in our study had a definite trigger, suggesting a possible relevance of avoiding/preventing exposure to a known precipitants. About one-third of cases of dystonic storm may occur unprovoked.[6] Temporizing therapy, supportive care, and antidystonia measures are the pillars recommended in the management of dystonic storm.[1] In the absence of a consensus statement, indication for mechanical ventilation needs to be based on institutional protocols along with objective dystonia grading scores. Based on our experience, we advocate an early and elective mechanical in dystonic storm to avoid bulbar, metabolic, and respiratory complication following high rates of sedation and anesthesia for control of the dystonia. Prevention of lung infection by following strict asepsis and critical care guidelines is vital.[1,7] Varying underlying etiologies mandate case-based treatment of underlying etiology. Yet, we achieved dystonia management using a uniform protocol. DSAP score is a simple, reproducible, and objective tool with good delineation of dystonic crisis.[8] This grading system in provides an objective benchmark for admission, management, and discharge of children. Other supportive care involves adequate hydration, nutrition pain relief, sedation, and airway protection.[9]

Figure 2 Dot Plot Depicting Intervention(s) in Case of Dystonic Storm

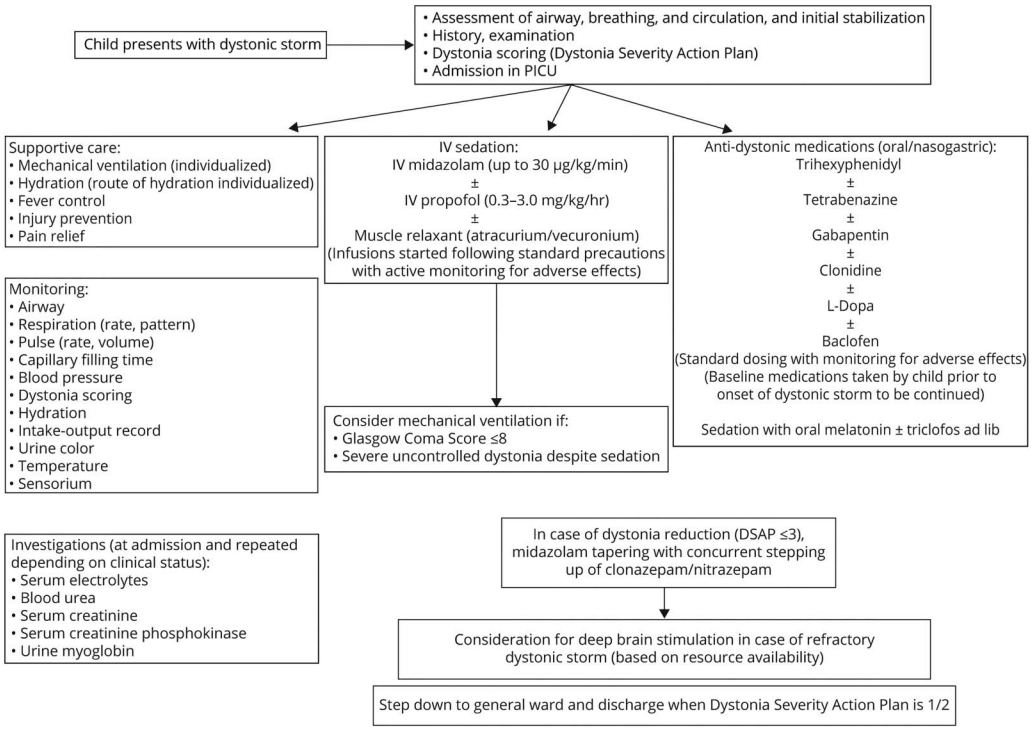


Numerical figures indicate number of cases. Suffix "R" in the diagnosis indicates that the particular case relapsed with dystonic storm at a later time point. AADC = aromatic L-amino acid decarboxylase; CP = cerebral palsy; IEM = inborn error of metabolism; PKAN = pantothenate kinase-associated neurodegeneration; SSPE = subacute sclerosing panencephalitis.

Drug therapy is reportedly effective in only upto 10% of cases of dystonic storm with midazolam being a key drug.[5] Our experience too suggests that pharmacologic management with injectable midazolam along with other drugs is the initial modality of choice to manage dystonic storm. Although greater percentage of children in our study had symptom cessation with pharmacotherapy, both the cohorts are not comparable due to multiple confounders. Symptomatic pharmacotherapy is as crucial and effective as definitive therapy for underlying etiology.[1,7,10] We suggest a combination polypharmacotherapy approach with multiple antidystonic drugs with differing mechanisms of action and benzodiazepines rather than a single agent uptitration strategy.[1,7]Trihexyphenidyl, tetrabenazine, and gabapentin were extensively used in our study. These drugs are commonly reported adjuncts for managing dystonic storm, although the percentage of usage is not reported in the literature.[11,12] Adequate sedation forms one of the cornerstones of management of a dystonic crisis since sleep is proven to be beneficial in decreasing dystonia.[1,13] Midazolam was used extensively in our cohort. This remains an effective sedative with short half-life, good cardiovascular profile with muscle relaxant properties, and an additional antidystonic effect. Efficacy is rapidly lost due to tolerance.[1,6,9] In the absence of IV clonidine, oral clonidine was used orally as an add-on therapy in our study with minimal adverse effects. Use of clonidine by oral, enteral, or IV route has been previously reported for dystonic crisis in children.[1,14] Alongside these agents, we relied on melatonin and triclophos for achieving adequate sedation. Nasogastric administration of these drugs was started even when child used to be on midazolam infusion. These medications had minimal adverse effects, were well tolerated in children, and enabled reduction of midazolam/clonazepam to which individual developed tolerance rapidly. The literature does not reveal use of these drugs specifically in dystonic crisis. Clonidine too has additional sedative effects.

Although deep brain stimulation (DBS) is a known effective modality for management of this entity, financial issues and

Figure 3 Algorithm for Dystonic Storm Management Followed in the Study Center



DSAP = Dystonia Severity Action Plan; PICU = pediatric intensive care unit.

requirement of experienced neurosurgery team may be challenges in DBS, especially in resource-constrained settings.[5,15–17] In our study, drug therapy was successful in all cases. Globus pallidus interna-DBS was performed in 1 child after resolution of symptoms. The child remains under follow-up with dystonia grade DSAP 1–2. Intrathecal baclofen is also effective in treating dystonic storm in both primary and secondary dystonia.[14,18] We did not prescribe this in view of the high cost and invasiveness entailed by this therapy.

Dystonic storm can be potentially fatal with multiple complications.[19] Multidrug therapy, adequate supportive care, and close monitoring were the keys in prevention of complications in our study. In our study, mean durations of hospitalization of children with neurologic conditions other than CP were significantly longer than those with CP. There is a ground to study possible correlation between treatment refractoriness of dystonic storm and underlying etiology. Relapse is common in cases with dystonia who present with dystonic storm.[7,17] Ensuring drug compliance, parental education, close follow-up, and early management of infections may be some strategies to minimize relapses. The simplistic model, sizeable patient population, and uniform protocolized drug usage policy are the strengths of the study.

Table 3 Details of Relapses in Children Postdischarge After Initial Hospitalization

Diagnosis	Sex	No. of relapses (R)	Precipitant of relapse	Complications (if any)
Mixed CP	Female	1	Unknown	None
Mixed CP	Female	3	R1: upper respiratory tract infection; R2: fever without focus; R3: unknown	Rhabdomyolysis (no renal failure)
Glutaric aciduria	Female	3	R1: missing trihexyphenidyl; R2: diarrhea; R3: missing trihexyphenidyl	R2 associated with regression of milestones
Idiopathic	Female	1	Unknown	None
Sequelae of Japanese encephalitis	Male	1	Unknown	None
Postencephalitic sequelae	Female	1	Unknown	None

Abbreviation: CP = cerebral palsy; R1 = relapse 1; R2 = relapse 2; R3 = relapse 3.

TAKE-HOME POINTS

- Pediatric dystonic storm occurs secondary to multiple etiologies.
- CP is the commonest etiology of pediatric dystonic storm.
- The trigger of dystonic storm may be identified in only about 50% of children.
- The core management tools of dystonic storm include meticulous assessment and monitoring, titrated polypharmacotherapy, adequate sedation, and balanced approach to elective mechanical ventilation.
- Dystonic storm relapses mostly due to preventable factors.

Possibility of referral bias and the retrospective study model are the inherent limitations of the study. The standard practices followed in the study center may not be practiced across all health care facilities due to nonavailability of pediatric neurology services and resource limitations, and hence, outcomes in the community setting may differ.

To conclude, in this retrospective, hospital-based study, fresh-onset dystonic storm occurred secondary to multiple varied etiologies with an identifiable trigger in only 50%. Children on follow-up for dystonic and mixed CP comprised a significant proportion of cases that had exacerbation of symptoms secondary to a triggering event. Objective DSAP scoring, titrated dosing of midazolam infusion, balanced approach to elective mechanical ventilation, and concurrent polypharmacotherapy were successful in safe management of this condition with minimal complications. Rates of rhabdomyolysis were insignificant. Adequate sedation helped in amelioration of symptoms and enabled early taper of benzodiazepines. Relapse occurred in 26% cases mostly due to preventable factors.

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Disclosure

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Appendix. Authors

Name	Location	Contribution
Jyotindra Narayan Goswami, MD, DNB, DM, MNAMS	New Delhi, India	Study conceptualization, data collection, data analysis, literature review, draft manuscript preparation, and approval of final manuscript
Shuvendu Roy, MD	Kolkata, India	Data collection, data analysis, draft manuscript preparation, and approval of the final manuscript
Saroj Kumar Patnaik, MD	Bangalore, India	Study conceptualization, supervision of the study, data collection, data analysis, and approval of final manuscript

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