Movement Disorder Emergencies

Dr Juzar Hooker

Consulting Neurologist

Aga Khan University Hospital, Nairobi

We shall discuss

- What is a movement disorder emergency?
- Frequency?
- Classification
- Approach
- Specific types

Movement disorder emergency?

“any neurological disorder, evolving acutely or subacutely, in which the clinical presentation is dominated by a primary movement disorder, and in which failure to accurately diagnose and manage the patient may result in significant morbidity or even mortality”

Fahn and Frucht 2002

Original Article

Spectrum of Movement Disorder Emergencies in a Tertiary Care Center in India: A Prospective Observational Study

Abhishek P. Bhoyar, Rohan Mahale, Nitish Kamble, Vikram Holla, Pramod Kumar Pal, Ravi Yadav
Department of Neurology, National Institute of Mental Health and Neurosciences, Bengaluru, Karnataka, India

2 year study 2019-2021

71 patients

65 hyperkinetic, 6 hypokinetic

Chorea 59%>dystonia>myoclonus (dystonia> in children)

Hyperglycaemia>stroke>autoimmune

Bhoyar et al 2022

Movement Disorder Emergencies

Hypokinetic

Acute parkinsonism
Neuroleptic malignant syndrome
Parkinsonism-hyperpyrexia syndrome

Catatonia

Acute exacerbation stiff person syndrome spectrum disorder

Hyperkinetic

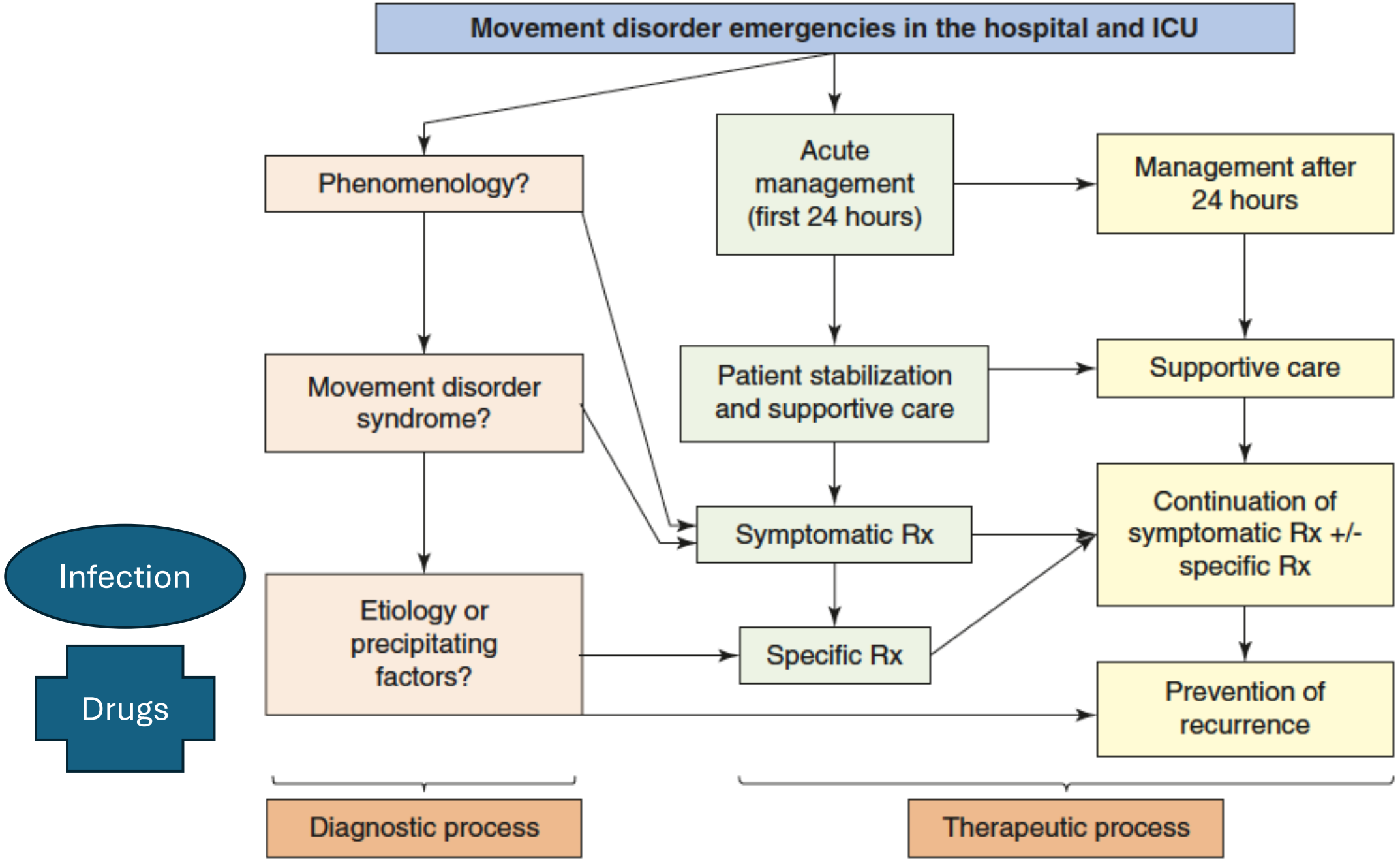
Status dystonicus/dystonic storm
Acute dystonic reaction

Serotonin syndrome

Myoclonic status

Choreic storm

Tic storm



Infection

Drugs

Modified from: Frucht S (Ed.) 2022. Movement Disorder Emergencies

1. Phenomenology	2. Syndrome	3. Etiology/Precipitating factor
Hypokinetic		
Parkinsonism	Neuroleptic malignant syndrome	Neuroleptics or other dopamine receptor blocking agents
	Parkinsonism-hyperpyrexia syndrome[1]	Abrupt discontinuation of levodopa
	Acute parkinsonism	CNS infection such as viral encephalitis, prion; drugs
Catatonia	Malignant catatonia	Underlying psychiatric disorders such as depression or schizophrenia
Stiffness	Acute severe exacerbation of spasms and rigidity/status spasticus in SPSD	Natural history of the diseases; can be triggered by intercurrent illness such as infection

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Hyperkinetic		
Dystonia	Dystonic storm	Infection, abrupt discontinuation of anti-dystonic treatment such as baclofen or anticholinergics
	Acute dystonic reaction	Neuroleptics or other dopamine receptor blocking agents
Myoclonus + tremor (also with rigidity)	Serotonin syndrome[2]	Serotonergic agents (dose-dependent)
Chorea	Choreic storm	Underlying choreic disorders; can be triggered by intercurrent illness such as infection
Myoclonus	Myoclonic status	Underlying myoclonic disorders such as posthypoxic myoclonus, not responsive to medical treatment
Tic	Tic status	Exacerbation of an underlying tic disorder

Frucht S (Ed.) 2022. Movement Disorder Emergencies

MD emergency syndrome	Main phenomenology	Co-existing features	Fever	Other autonomic features	Altered mental status
Neuroleptic malignant syndrome[a]	Parkinsonism including rigidity		++	++	++
Serotonin syndrome	Myoclonus, shivering-like movements	Rigidity, hyperreflexia	+	++	++
Malignant catatonia	Catatonia		+	+	++
Acute severe exacerbation of spasms and rigidity/ status spasticus in SPSD	Stiffness, muscle spasms, and rigidity		+	++	+/++
Acute dystonic reaction	Dystonia, typically retrocollis and lower cranial dystonia	Can co-occur with oculogyric crisis	+/–	+/–	–
Dystonic storm	Dystonia		+	+	–
Choreic storm	Chorea		+/–	+/–	–
Myoclonic status	Myoclonus		+	+	–
Tic status	Tic		–	–	–

1. Acute management in the first 24 hours
Maintain ABC (Airway, Breathing and Circulation)
Early ICU transfer: Be aggressive and proactive
Secure IV access
Aggressive IV fluid hydration
Temperature reduction measures, e.g., acetaminophen or cooling blanket if >40 °C
Treatment of autonomic instability such as hypertension, tachycardia
Assess and consider needs for ventilatory support
Close monitoring of vital signs and other parameters such as urine output
Obtain necessary lab tests, e.g., CBC, electrolytes, BUN, Cr, CK, UA, ABG, infectious work-up, but these should not delay other steps
Symptomatic therapies of movement disorders
Selection is based on the phenomenology and movement disorder emergency syndromes (see Table 1.1)
IV sedatives/anesthetics, if needed, e.g., benzodiazepines, propofol, barbiturates
Specific therapies: Address and treat underlying etiology and precipitating factors
2. Management after 24 hours
Continue supportive care and symptomatic therapies
Prevent recurrence or relapses

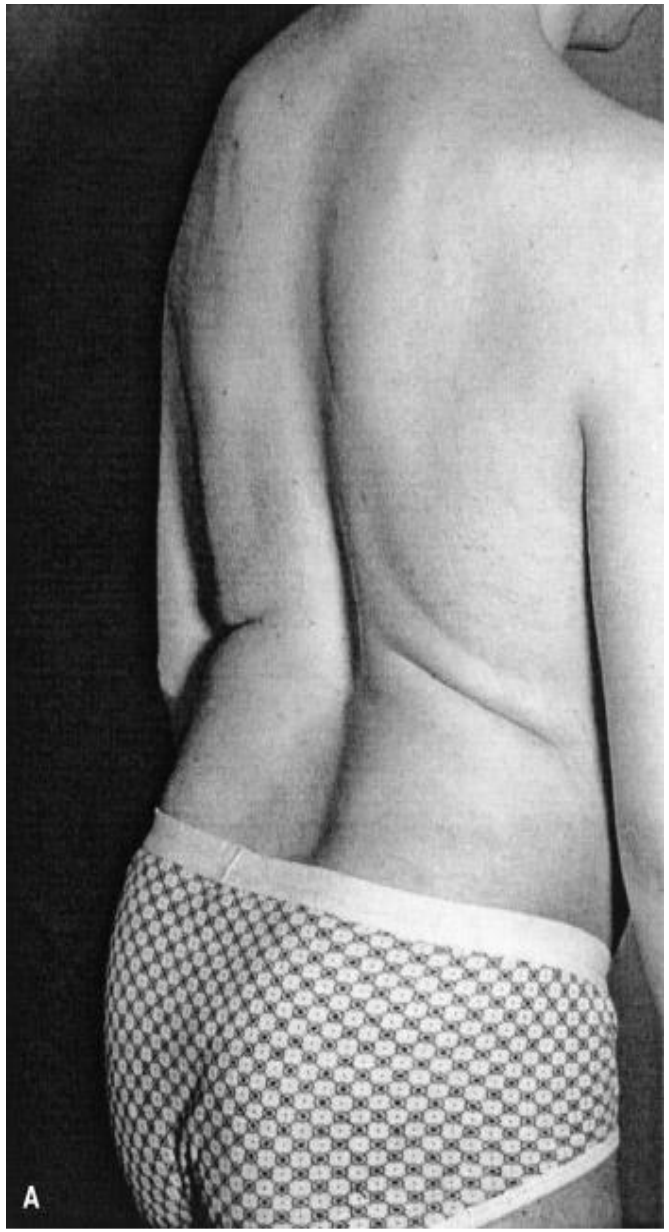
Movement disorder emergency syndrome	Symptomatic Rx	Specific Rx
Neuroleptic malignant syndrome	Dopamine agonists, e.g., bromocriptine; Dantrolene	Discontinue neuroleptics; avoid restarting at least in the next 2 weeks
Serotonin syndrome	Cyproheptadine, propranolol	Discontinue serotonergic agents
Malignant catatonia	BZDs such as lorazepam	Treat underlying psychiatric disorders; consider electroconvulsive therapy (ECT)
Acute severe exacerbation of spasms and rigidity/status spasticus in SPSD	BZDs, baclofen, anesthetic agents	Treat precipitating factors such as infection
Acute dystonic reaction	IV anticholinergics or antihistamines, BZDs may be an alternative	Discontinue neuroleptics
Dystonic storm	BZDs, baclofen, anticholinergics, sedative/anesthetic agents; DBS can be considered in DYT1, DYT6, and tardive dystonia	Treat underlying precipitating factors; resume baclofen in case of baclofen withdraw
Choreic storm	High-potency neuroleptics, tetrabenazine, sedative/anesthetic agents	Treat underlying etiologies of chorea such as blood sugar control in non-ketotic hyperglycemia
Myoclonic status	Valproate, levetiracetam, BZDs; DBS can be considered in posthypoxic myoclonus	Treat underlying precipitating factors, if any
Tic status	High potency neuroleptics, tetrabenazine, BZD, clonidine	Treat underlying precipitating factors, if any

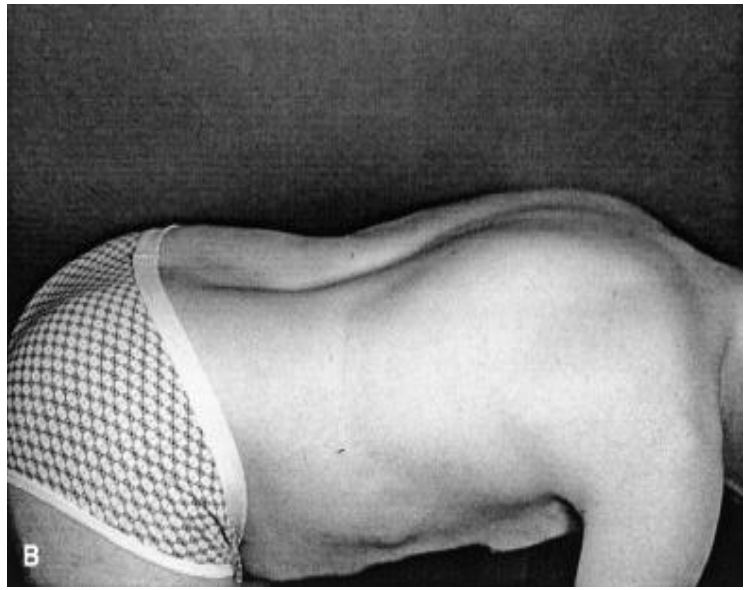
Medication	Route[a]	Dosage	Dosing interval	Note
Bromocriptine	PO/ NG	2.5–5 mg/dose	q 8 h	If not available, other DAs can be considered as an alternative
Dantrolene	IV	1–2.5 mg/kg/dose	q 6 h	
Cyproheptadine	PO/ NG	2–12 mg/dose	q 2 h	Start with 12 mg, followed by 2 mg q 2 h
Benztropine	IV/IM	1–2 mg/dose	Single dose	
Diphenhydramine	IV	25–50 mg/dose	Single dose	
Baclofen	PO/ NG	15–120 mg/d	TID	Intrathecal form should *not* be injected intravenously
Lorazepam[b]	IV/IM	1–4 mg/dose	q 5 min to 4–6 h	Dosing interval depends on indications[c]
Diazepam[b]	IV/IM	5–10 mg/dose	q 8 min to 8 h	Dosing interval depends on indications[c]
Midazolam[b]	IV	Initiation 0.01–0.05 mg/kg; maintenance 0.02–0.1 mg/kg/h	Continuous infusion	
Propofol	IV	Initiation 0.3 mg/kg/h; maintenance 0.3–3 mg/kg/h	Continuous infusion	
Pentobarbital	IV	Loading 1 mg/kg, then 1–3 mg/kg/h (maximum 5 mg/kg/h)	Continuous infusion	

Table 1.6 Pearls and pitfalls in the management of movement disorder emergencies in the hospital and intensive care unit

Be aggressive, intervene early. Do not exercise a "wait-and-see" approach.
Consider early ICU admission.
General ICU care including vital sign monitoring, IV fluid hydration, temperature reduction measures is paramount.
Aggressive IV fluid hydration to prevent renal failure, especially when at risk for rhabdomyolysis and myoglobinuria.
Close monitoring is required.
An IV route of medication administration is preferred in the first 24 hours.
Maintain with oral symptomatic medication(s) after acute treatment with IV medications; early discontinuation of symptomatic therapies may lead to recurrence.
Be cautious about movement disorder emergency mimics such as pseudodystonic emergencies and sepsis (mimicking neuroleptic malignant syndrome or malignant hyperthermia).

Stiffness





Meinck and Thompson 2002

Pathophysiology	Causes
Vascular	Spinal arteriovenous malformations
Infectious	Tetanus
	Rabies
Drug-induced	Neuroleptic malignant syndrome
	Parkinsonism–hyperpyrexia syndrome
	Serotonin syndrome
Toxic	Strychnine
Metabolic	Hypocalcemia
Inherited	Hyperekplexia
	Malignant hyperthermia
Autoimmune	Stiff man syndrome
Psychiatric	Lethal catatonia

Kipps et al 2005

Stiff person syndrome spectrum disorder: the antibodies

- Anti-glutamic acid decarboxylase antibodies (GAD) (SPS)
- Anti-amphiphysin antibodies (paraneoplastic) (SPS)
- Anti-glycine receptor antibodies (PERM)
- Anti-gephyrin antibodies (PERM)
- Anti-Ri antibodies (paraneoplastic) (PERM)

Modified from: Frucht S (Ed.) 2022. Movement Disorder Emergencies

Stiff person syndrome spectrum disorder: management

Benzodiazepines: diazepam, clonazepam
GABA analog: baclofen
Centrally acting antiadrenergic: tizanidine, clonidine
Anticonvulsants: gabapentin, valproate
Botulinum toxin injections for focal rigidity, spasm
Immunotherapy: PERM, SPS

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Parkinsonism

Pathophysiology	Causes
Vascular and structural	Basal ganglia stroke (esp. involving globus pallidus)
	Midbrain lesions
	Hydrocephalus
Infectious	Encephalitis lethargica
	Other viral encephalitis (esp. Japanese B)
	Mycoplasma
Drug induced	Parkinsonism–hyperpyrexia syndrome
	Chemotherapy
	Amphotericin B
Toxic	Carbon monoxide
	Methanol
	Cyanide
	Organophosphate poisoning
	MPTP
Metabolic	Central pontine myelinosis
Inherited	Rapid-onset dystonia–parkinsonism
Psychiatric	Neuroleptic-induced

Kipps et al 2005

Infectious
Post-infectious
Autoimmune/paraneoplastic Systemic lupus erythematosus LGI1 antibodies IgLON5 antibodies Dopamine 2 receptor antibodies Ma2 antibodies Ri antibodies
Medication “Typical” side effects of anti-dopamine drugs Idiosyncratic effects Neuroleptic malignant syndrome Serotonin syndrome Chemotherapeutic drugs

Frucht S (Ed.) 2022. Movement Disorder Emergencies

- Toxic
  - Carbon monoxide
  - Cadmium
  - MPTP
  - Ethanol withdrawal
  - Ethylene oxide
  - Methanol
  - Disulfiram
  - Bone marrow transplantation
  - Organophosphate exposure
- Structural
  - Stroke
  - Subdural hematoma
  - Central and extra pontine myelinolysis
  - Tumor
  - Hydrocephalus

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Psychiatric

- Catatonia
- Conversion
- Obsessive-compulsive disorder (obsessional slowness)
- Malingering

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Management of Acute Parkinsonism

- Withdrawal of offending agents
- Anticholinergic medication (diphenhydramine, trihexyphenidyl)
- Dopamine agonist
  - Neuroleptic maligant syndrome
- Levodopa
  - Caution with postencephalitic parkinsonism (levodopa sensitivity)

Management of Acute Parkinsonism

- Lesioning

- Deep Brain Stimulation (DBS)

Emergencies in Parkinson’s disease

- Psychosis
- Medication withdrawal
- DBS failure and DBS related emergencies

Psychosis in PD

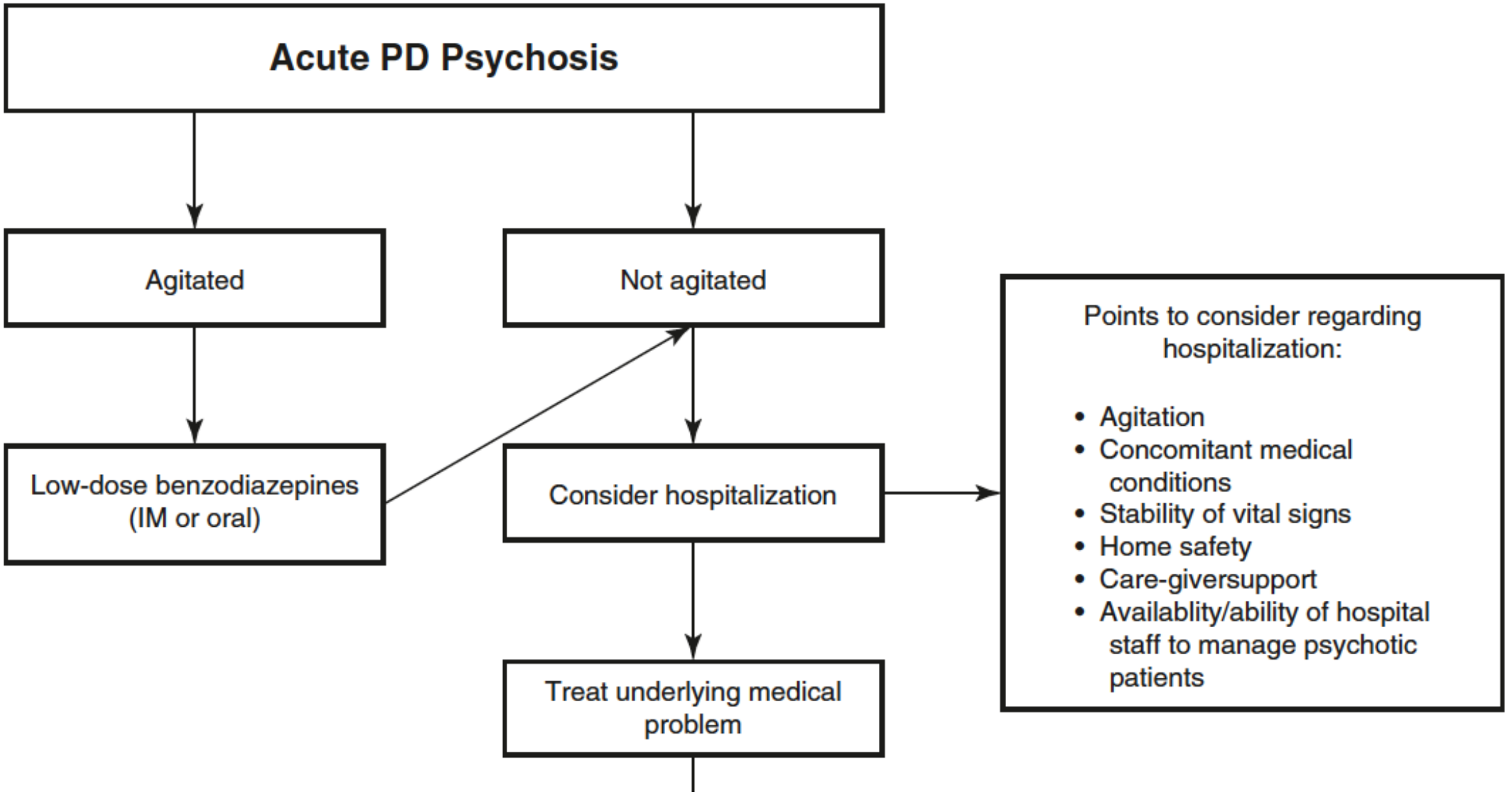
Table 17.1 Risk factors for PD psychosis

Older age
Older age at PD onset
Worse motor function, particularly axial function
Higher baseline levodopa equivalent dose
Advanced disease
Lower cognitive status
Concomitant dementia
Concomitant depression
Sleep disturbances (e.g., REM sleep behavior disorder)
Multiple medical problems
Visual perceptive disorders

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Differential diagnosis
P—Parkinson's disease medications
SY—Systemic illness
C—Centrally acting medication
H—Hepatic, renal, or other metabolic dysfunction
O—Overdose of medications or intoxication
S—Sensory deprivation (hearing, visual impairment)
I—Infection (urinary tract infection, pneumonia)
S—Structural lesions (stroke, subdural hematoma, intracranial hemorrhage, trauma)

Frucht S (Ed.) 2022. Movement Disorder Emergencies



Frucht S (Ed.) 2022. Movement Disorder Emergencies

Discontinue or reduce non-PD related medications with psychoactive (or CNS) properties

Titrate or eliminate:

- Anticholinergics
- Amantadine
- MAO-B inhibitor
- Dopamine agonist
- COMT-inhibitor
- Levodopa

Treat with antipsychotics (e.g., clozapine, pimavanserin, quetiapine)

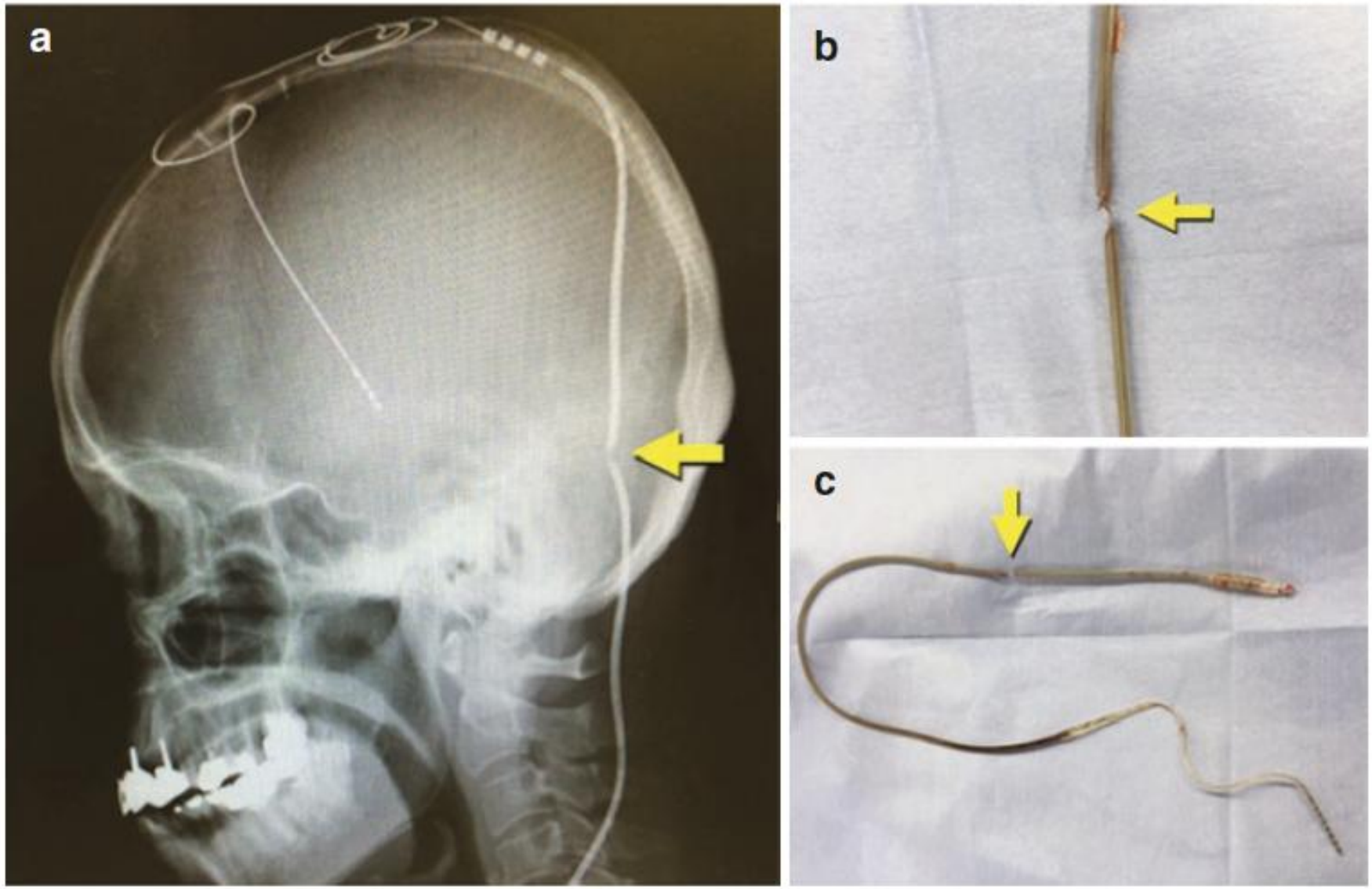
Modified from: Frucht S (Ed.) 2022. Movement Disorder Emergencies

DBS related emergencies

DBS failure

- Battery failure

- Lead fracture/hardware hitch



As shown in: Frucht S (Ed.) 2022. Movement Disorder Emergencies

Perioperative DBS related emergencies

Table 18.1 Management of DBS-related emergencies

Issues	Management
Intraoperative emergencies	
Intracerebral hemorrhage	If the hemorrhage is very large or has symptomatic mass effect, an emergent craniotomy may need to be performed
Intraventricular hemorrhage	Ventriculostomy, if necessary, for obstructive hydrocephalus
Subdural hematoma	Bur hole irrigation should be performed when the hematoma is symptomatic
Air embolus	Wax edges of the bur hole, occlude the bur hole with gel foam and saline, lower patient's head, jugular venous compression, administer oxygen
Dyskinetic storm	Sedative agents may be administered in select cases. Reducing the dopaminergic medication may help. In some cases, ICU care is necessary
Epileptic seizure	Sedative agents such as propofol or a benzodiazepine such as diazepam should be administered immediately

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Early postoperative emergencies (<2–4 weeks)	
Venous infarction	Conservative supportive therapy is usually all that is necessary. An emergent craniotomy may be performed if hemorrhage is life-threatening
Myocardial infarction	Do not ignore chest pain in a patient who has just had a subclavicular IPG placed. Immediate diagnosis by 12-lead electrocardiogram and laboratory investigation, and cardiology consult should be performed
Neuroleptic malignant syndrome	IV fluid and L-dopa should be administered immediately. If necessary nasogastric tube should be placed for patients with difficulty in oral intake. ICU care is necessary. Administering dantrolene is an option

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Issues	Management
Behavioral/cognitive issues	Identify and treat the underlying issues (e.g., UTI and pneumonia). Selective dopamine blockers (e.g., clozapine, quetiapine) may be used, but nonselective blockers should be avoided if possible. Use a one-to-one sitter to avoid secondary injury, e.g., from falling
Infection-UTI/ pneumonia	Hydration and appropriate antibiotics. Care should be taken to adjust PD medications as levels may be altered by antibiotics. Surgical debridement or removal of hardware as necessary

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Postoperative emergencies (≥2–4 weeks)	
Suicide ideation/ attempt	Admit the patient to the hospital for multi/interdisciplinary care, and treat underlying cause. May need both medication adjustment and programming. Check lead location
Severe depression	Behavioral therapy, counseling, medication adjustment, and/or stimulation adjustment. Check lead location. Consider admission for multi/interdisciplinary management
Superficial wound infection	Administration of antibiotics or surgical revision should be considered before development of deep infection
Infection-lead	The lead should be removed and appropriate antibiotics should be administered
Infection-IPG	The IPG and usually the extension cable should be removed and appropriate antibiotics should be administered
Lead migration	Lead replacement, or surgical alteration of lead position
Lead fracture	Lead replacement, if an appropriate candidate
Lead electrical short	Lead replacement, or potentially reprogramming at a different contact
IPG malfunction	IPG replacement, manage potential rebound symptoms
Accidental on/off	Turn on the IPG. Educate the patient and the family so they can use on/ off devices
Symptom rebound (motor and/or non-motor)	DBS hardware workup including impedance check, battery check, X-ray study, and assess for tolerance
Intracranial cyst	Steroids may be tried in mild cases but treatment usually requires removal of DBS lead

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Multiple system atrophy

- Stridor
- Mortality risk
- Mechanism
  - Abductor dysfunction
  - Adductor dystonia

Management

- CPAP

- Tracheostomy

Neuroleptic malignant syndrome

What is it?

Hyperthermia/fever
+
Movement Disorder/Rigidity
+
Altered Mental Status
+
Autonomic instability

Dopamine receptor antagonist exposure

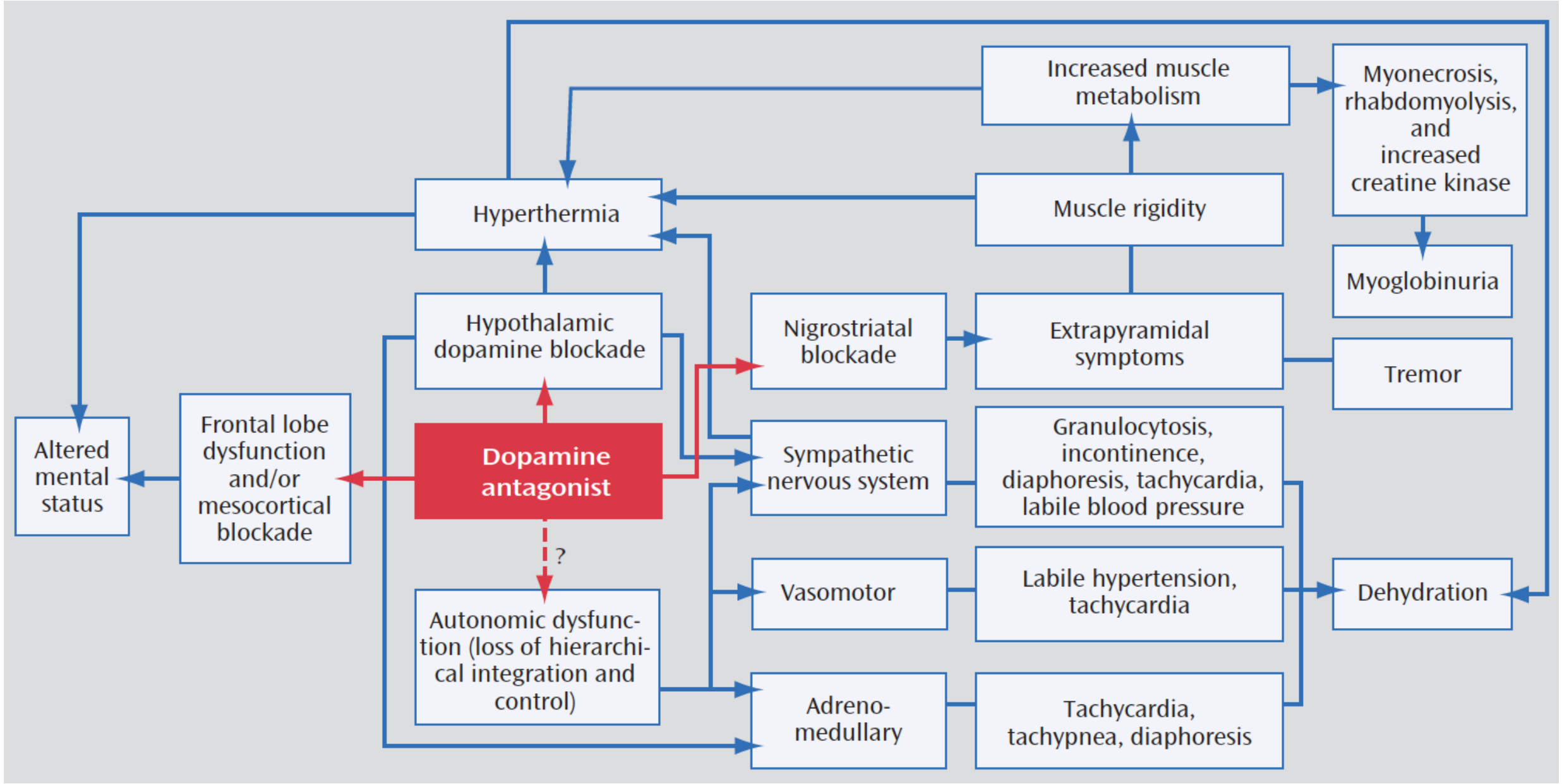
The impact

- Dopamine receptor antagonist use
- 1:5000
- 10% mortality
- Significant medium term morbidity

Gurrera et al 2007; Stubner et al 2004; Spivak et al 2000

Risk factors

- Young adults
- M>F 3:2
- Non-white
- Anti NMDA receptor antibody associated autoimmune encephalitis



Strawn et al 2007

Table 6.1 Clinical features of NMS

Administration of dopamine-receptor blocking agents
Signs and symptoms
Hyperthermia (>38 °C)
Muscle rigidity ± cogwheeling
Tremor and myoclonus
Mental status changes (stupor, mutism, and delirium)
Autonomic instability (tachycardia and labile blood pressure)
Tachypnea and dyspnea
Diaphoresis, sialorrhea, and incontinence
Dysarthria and dysphagia
Associated laboratory findings
Muscle enzyme elevations (CK, LDH, transaminases, and aldolase), myoglobinuria, leukocytosis, metabolic acidosis, hypoxia, low serum iron, elevated serum catecholamines, and slowing on EEG
Complications
Cardiorespiratory arrest, acute renal failure, rhabdomyolysis, pulmonary emboli, aspiration pneumonia, disseminated intravascular coagulation, limb contractures, and ischemic brain damage
Exclusion of other central, systemic, and toxic causes of hyperthermia

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Table 6
Clinical tetrad and associated features of neuroleptic malignant syndrome

Clinical Tetrad	Associated Features
Hyperthermia	Elevated creatine kinase
>38°C (87%)	Proteinuria
>40°C (40%)	Myoglobinuria
Autonomic Disturbances	Leukocytosis with PMN predominance
Tachycardia	Transaminitis
Diaphoresis	Hypomagnesemia
Labile blood pressure	Hypocalcemia
Extrapyramidal Signs	Reduced ferritin
Rigidity	Severity Scale
Bradykinesia	Mild NMS: mild rigidity, confusion, temperature ≤38°C, and tachycardia ≤100 bpm
Dystonia	Moderate NMS: stupor, catatonia, hyperpyrexia 38–40°C, or tachycardia 100–120 bmp
Tremor	Severe NMS: severe rigidity, catatonia, coma, hyperthermia ≥40°C, or tachycardia ≥120 bpm
Alteration of Mentation	
Agitation	
Inattention	
Confusion	
Depressed mental status	
Catatonia	
Coma	

Radmard 2025

TABLE 1. Differential Diagnosis of Neuroleptic Malignant Syndrome

Infectious
Meningitis or encephalitis
Postinfectious encephalomyelitis syndrome
Brain abscess
Sepsis
Psychiatric or neurological
Idiopathic malignant catatonia
Agitated delirium
Benign extrapyramidal side effects
Nonconvulsive status epilepticus
Structural lesions, particularly involving the midbrain
Toxic or pharmacological
Anticholinergic delirium
Salicylate poisoning
Malignant hyperthermia (inhalational anesthetics, succinylcholine)
Serotonin syndrome (monoamine oxidase inhibitors, triptans, linezolid)
Substances of abuse (amphetamines, hallucinogens)
Withdrawal from dopamine agonists, baclofen, sedative-hypnotics, and alcohol
Endocrine
Thyrotoxicosis
Pheochromocytoma
Environmental
Heatstroke

Strawn et al 2007

Table 6.2 International Expert Consensus NMS Diagnostic Criteria [75]

Diagnostic criterion	Priority score
Exposure to dopamine antagonist, or dopamine agonist withdrawal, within past 72 h	20
Hyperthermia (>100.4 °F or >38.0 °C on at least two occasions, measured orally)	18
Rigidity	17
Mental status alteration (reduced or fluctuating level of consciousness)	13
CK elevation (at least four times upper limit of normal)	10
Sympathetic nervous system lability, defined as at least two of the following:	10
Blood pressure elevation (systolic or diastolic ≥25% above baseline)	
Blood pressure fluctuation (≥20 mmHg diastolic change or ≥25 mmHg systolic change within 24 h)	
Diaphoresis	
Urinary incontinence	
Hypermetabolism, defined as heart rate increase (≥25% above baseline) and respiratory rate increase (≥50% above baseline)	5
Negative work-up for infectious, toxic, metabolic, and neurologic causes	7
Total	100

>74

=

NMS

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Table 6.3 Proposed treatment algorithm for NMS [9]

Woodbury stage [88]	Clinical presentation	Supportive care	First-line interventions	Second-line interventions
Stage I: drug-induced parkinsonism	Rigidity; tremor	Discontinue, reduce or switch antipsychotics	Anticholinergic agents	
Stage II: drug-induced catatonia	Rigidity; mutism; stupor	Discontinue, reduce, or switch antipsychotics	Lorazepam (1–2 mg i.m. or i.v. every 4–6 h)	
Stage III: mild or early NMS	Mild rigidity; catatonia or confusion; temperature >38 °C (100.4 °F); heart rate >100 bpm	Discontinue antipsychotics; carefully monitor for progression; correct risk factors	Lorazepam (1–2 mg i.m. or i.v. every 4–6 h)	

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Stage IV: moderate NMS	Moderate rigidity; catatonia or confusion; temperature 38–40 °C (100.4–104 °F); heart rate 100–120 bpm	Discontinue antipsychotics, manage fluids, initiate cooling measures, correct risk factors, provide intensive care	Lorazepam (1–2 mg i.m. or i.v. every 4–6 h), bromocriptine (2.5–5 mg p.o. or by nasogastric [NG] tube every 8 h), or amantadine (100 mg p.o. or by NG tube every 8 h)	Consider electroconvulsive therapy (6–10 bilateral treatments)
Stage V: severe NMS	Severe rigidity; catatonia or coma; temperature >40 °C (104 °F); heart rate >120 bpm	Discontinue antipsychotics, manage fluids, initiate cooling measures, correct risk factors, provide intensive care	Dantrolene (1–2.5 mg/kg i.v. every 6 h. for 48 h), bromocriptine (2.5–5 mg p.o. or by NG tube every 8 h), or amantadine (100 mg p.o. or by NG tube every 8 h)	Consider electroconvulsive therapy (6–10 bilateral treatments)

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Parkinsonism-hyperpyrexia syndrome

What is it?

- Rare
- Potentially life threatening
- Patients with PD and other forms of parkinsonism
- Rapid ↓/stoppage dopaminergic medication or DBS
- Neuroleptic malignant syndrome like

Impact

- 1 day to 1 week of change in dopaminergic therapy
- Mortality 4%
- Long term sequelae 1/3

Precipitants

- Abrupt medication changes
- Medication non-adherence
- Coadministration of neuroleptics
- Dehydration
- Excessively hot temperatures
- Abruptly stopping deep-brain stimulation (DBS)
- Rapidly reducing antiparkinsonian therapy doses in patients on DBS

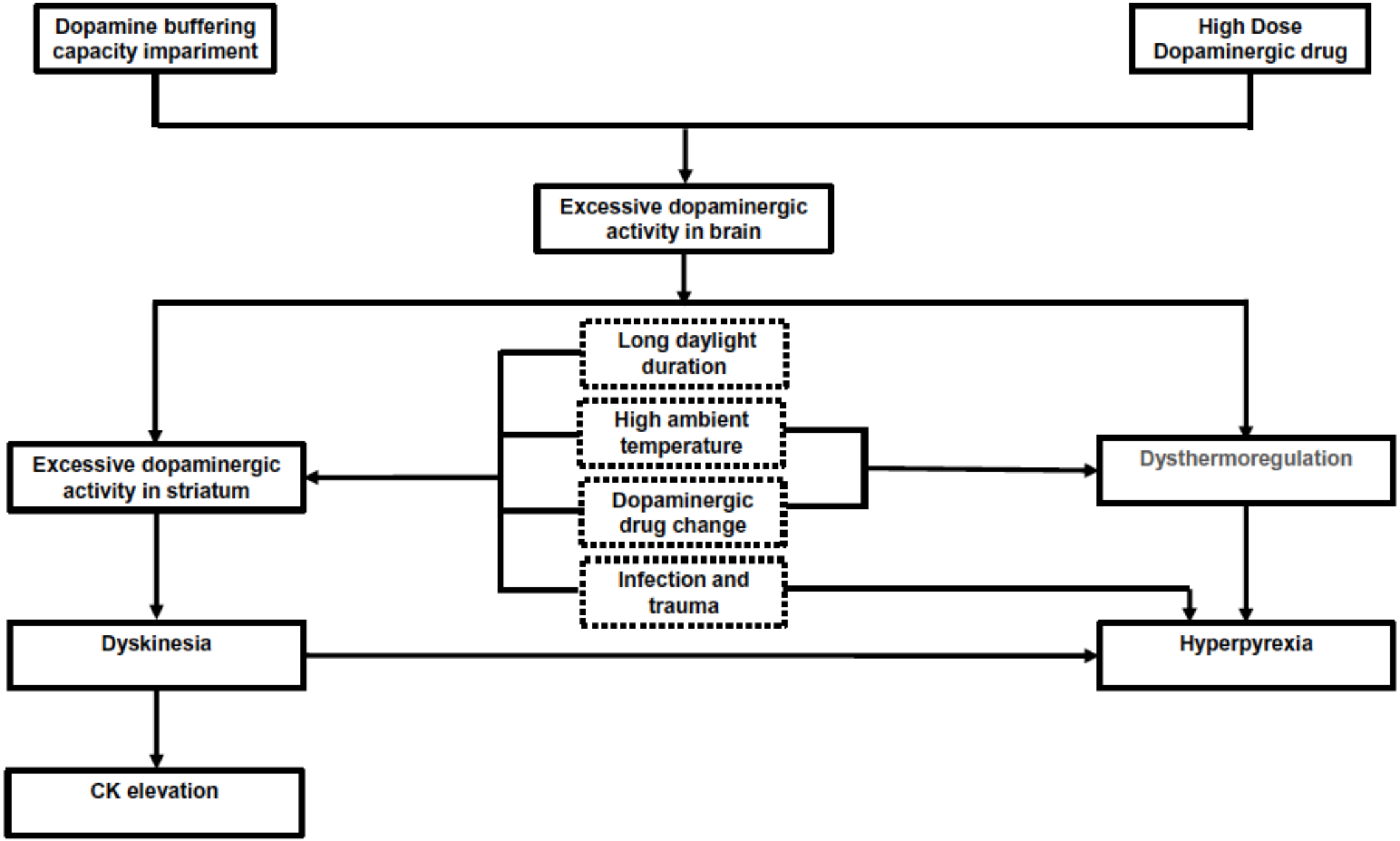
Recognition of the disorder
Verification of patients' medication regimen/compliance
Reintroduction of anti-parkinsonian medications
Supportive measures anti-pyretics/cooling blankets
Re-hydration
ICU monitoring/management (see text)
Clinical evaluation for possible co-morbid conditions
Bromocriptine 2.5 mg po t.i.d., titrated by 2.5 mg t.i.d./daily as necessary
Dantrolene sodium 10 mg/kg/d IV in divided doses (t.i.d./q.i.d.) as necessary
Apomorphine 2 mg SC every 3 h as necessary (if no nasogastric tube access, or use as adjunct)

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Dyskinesia-hyperpyrexia syndrome

Essential feature	Core feature	Supportive features
Severe and generalized dyskinesia	Hyperpyrexia	CK elevation or rhabdomyolysis Consciousness disturbance Autonomic dysfunction

Wang et al 2020



Wang et al 2020

Management

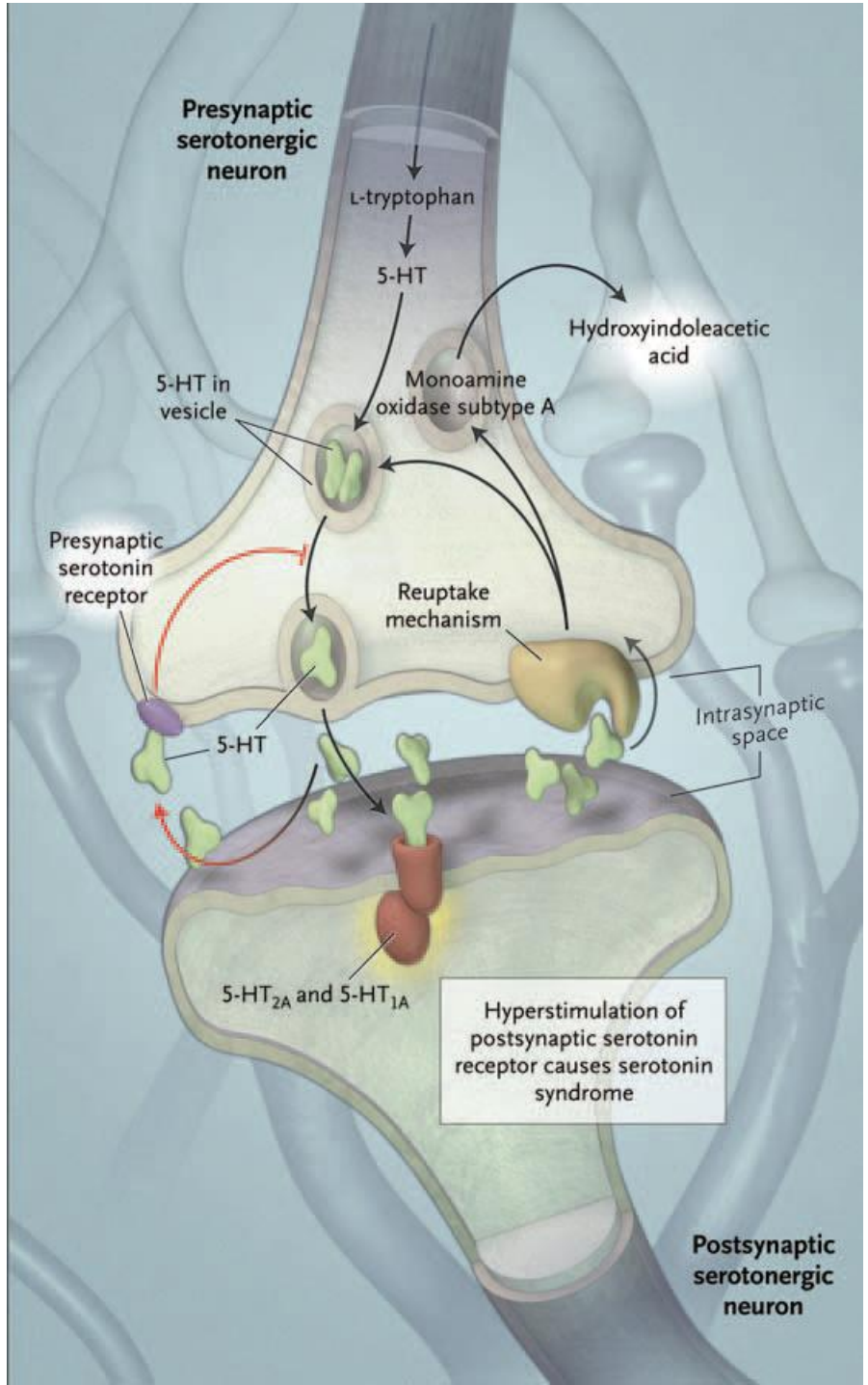
- Dopaminergic medication reduction
- Temperature reduction
- Fluids, support

Serotonin syndrome

What is it?

- Iatrogenic disorder arising from enhanced serotonergic activity mediated by brainstem 5HT1A receptors
- Increased central serotonin concentrations reduce central concentrations of dopamine, inducing a hypodopaminergic state similar to that in neuroleptic malignant syndrome
- Cognitive, autonomic and neuromuscular features

Gandhi et al 2020



Boyer and Shannon 2005

Table 8.1 Clinical manifestations of the serotonin syndrome [2]

Clinical manifestations	Features
Cognitive and behavioral	Confusion/disorientation
	Agitation/irritability
	Coma/unresponsiveness
	Hallucinations (visual and auditory)
Autonomic excitation	Hyperthermia
	Diaphoresis
	Sinus tachycardia
	Hypertension
	Dilated pupils
	Nausea
	Flushing
Neuromuscular features	Myoclonus (especially in the legs)
	Hyperreflexia (in the legs more than the arms)
	Muscle rigidity
	Restlessness/hyperactivity
	Tremor
	Ataxia
	Extensor plantar responses

Frucht S (Ed.) 2022. Movement Disorder Emergencies

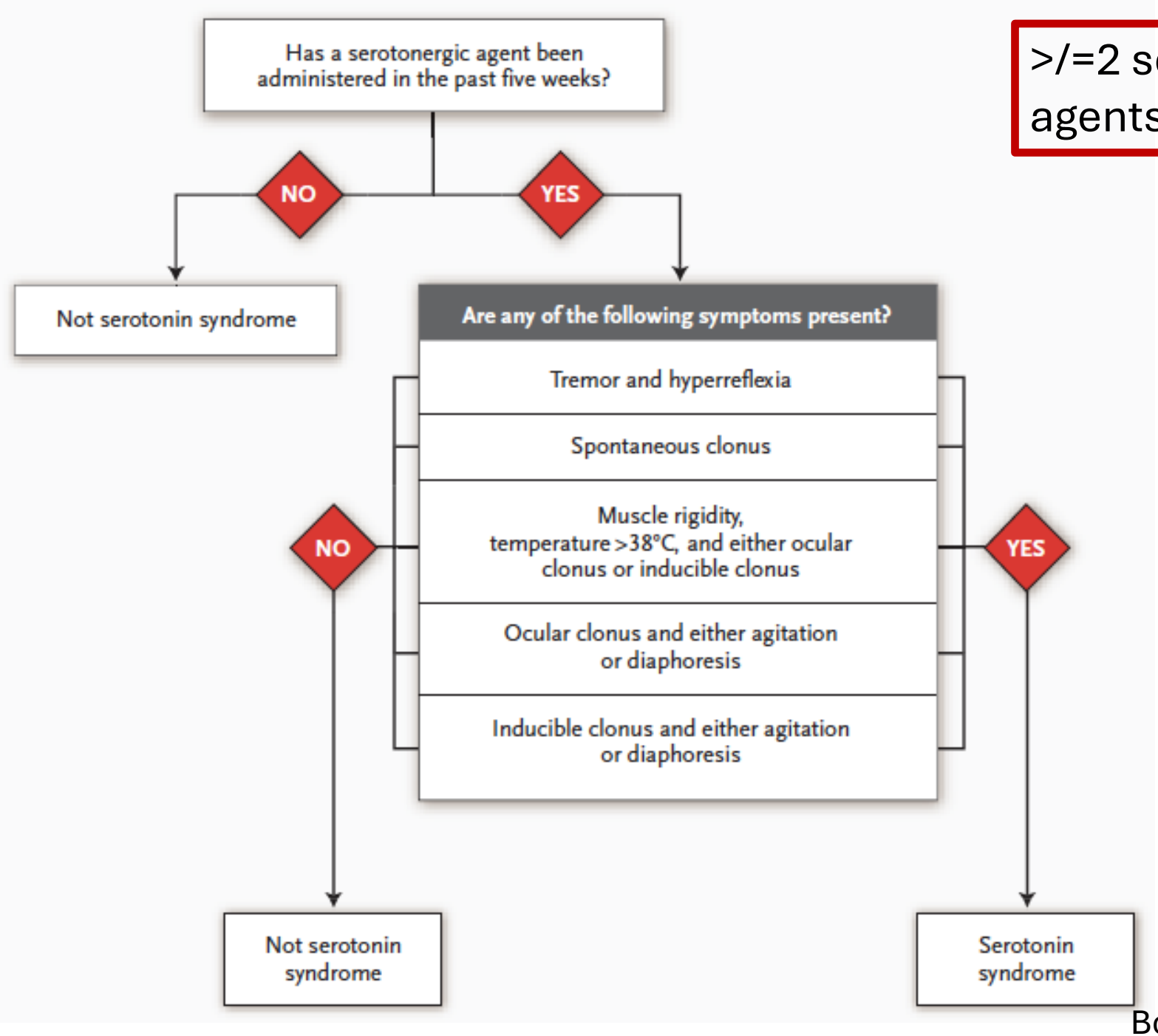
Table 1
Diagnosis of serotonin syndrome: the Hunter serotonin toxicity criteria

Use of at least one proserotonergic medication AND	At least ONE of the following clinical findings:
	Spontaneous clonus
	Inducible clonus AND agitation *or* diaphoresis
	Ocular clonus AND agitation *or* diaphoresis
	Inducible clonus AND increase muscle tone AND hyperthermia (temp >38oC)
	Ocular clonus AND increase muscle tone AND hyperthermia (temp >38oC)
	Tremor and hyperreflexia

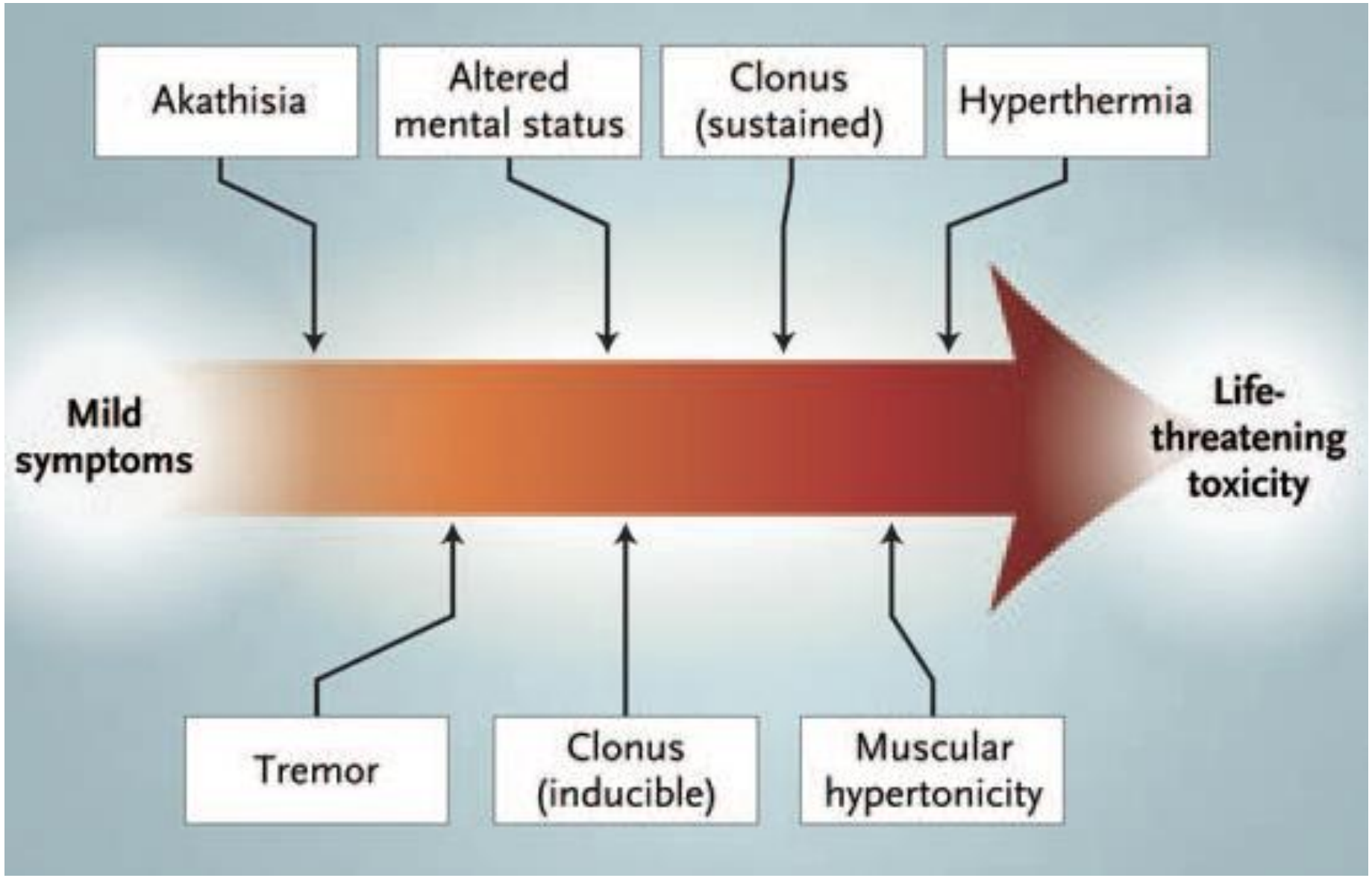
Adapted from Lerner DP, Tadevosyan A, Burns JD. Neurol Clin. 2020.

Apetauerova et al 2021

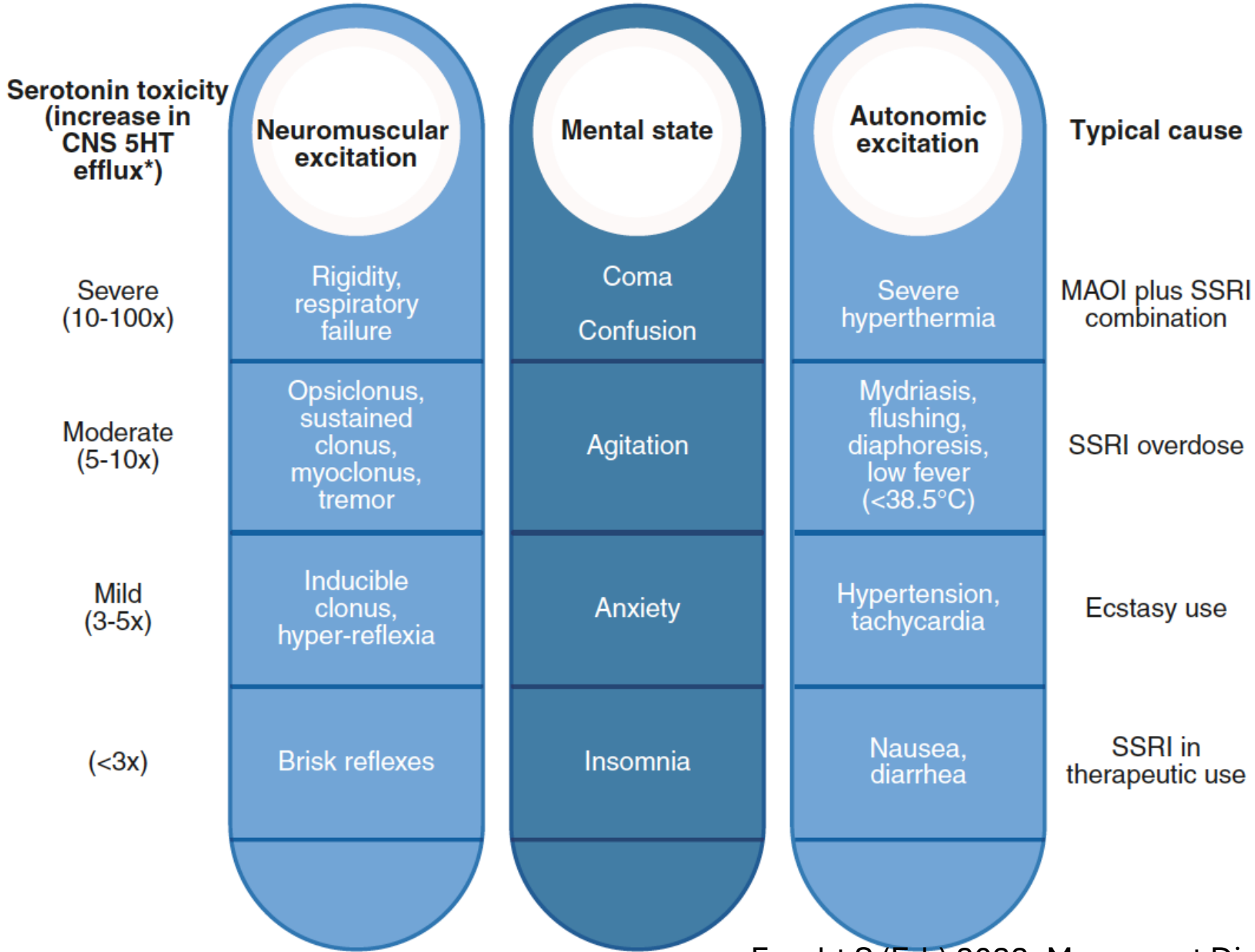
>/=2 serotoninergic agents/interacting



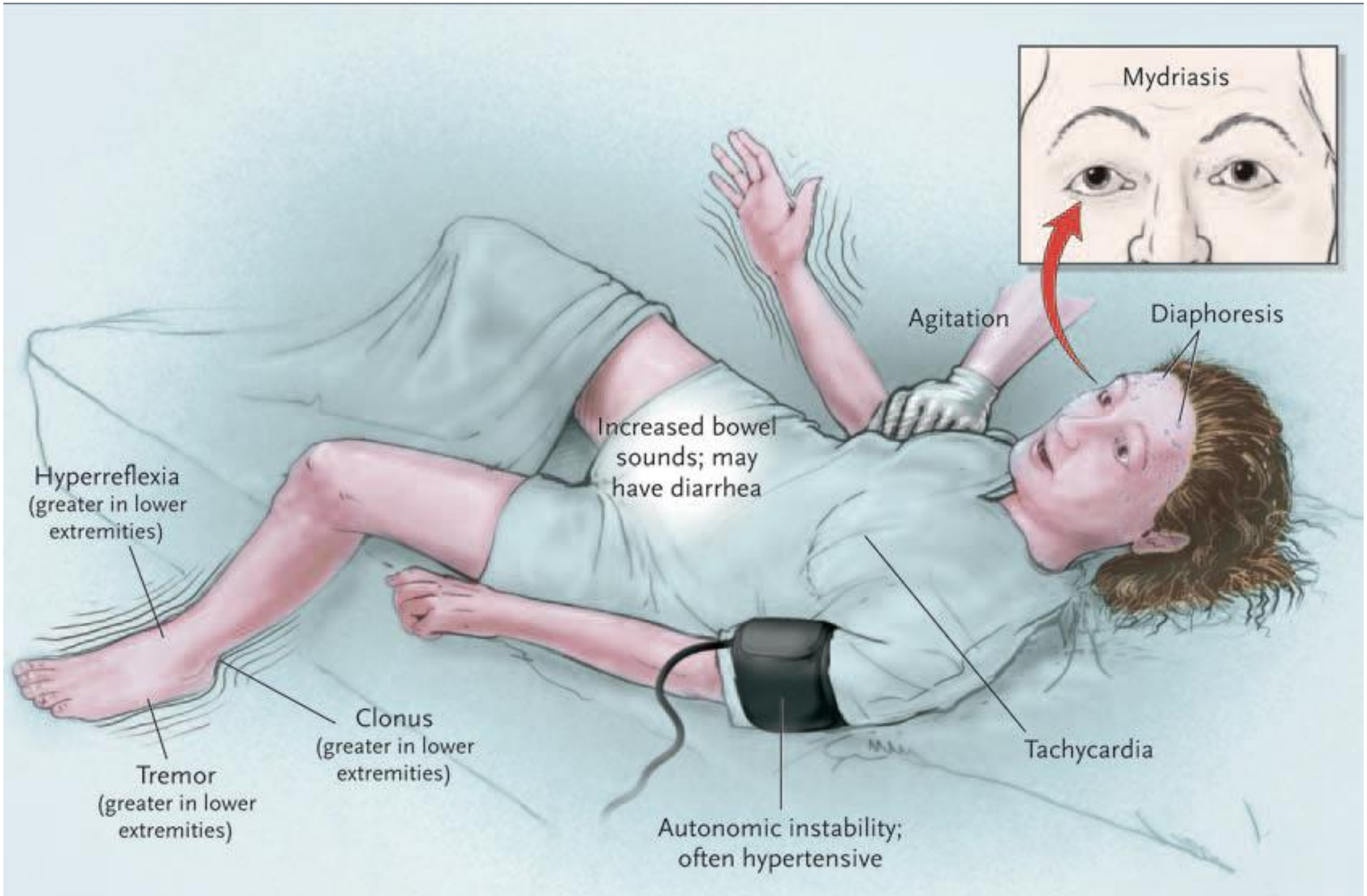
Boyer and Shannon 2005



Boyer and Shannon 2005



Frucht S (Ed.) 2022. Movement Disorder Emergencies



Boyer and Shannon 2005

- Neuroleptic malignant syndrome (NMS)
- Malignant hyperthermia
- Anticholinergic poisoning
- Heatstroke
- Serotonergic discontinuation syndrome
- Central hyperthermia
- Cerebral vasculitis
- Thyroid storm
- Delirium
- Delirium tremens
- Sympathomimetic overdose
- Meningitis
- Encephalitis
- Tetanus
- Alcohol or drug withdrawal
- Nonconvulsive seizures
- Stiff person syndrome

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Table 5
Commonalities and distinctions between presentation and management of serotonin syndrome and neuroleptic malignant syndrome

	Serotonin Syndrome	Neuroleptic Malignant Syndrome
Onset	Acute	Gradual
Hyperthermia	+	+
Dysautonomia	+	+
Depressed mentation	+	+
Opsoclonus	+	-
Pupils	Dilated	Normal
Rigidity	+	+
Tremor	+	-
Hyperreflexia	+	-
Clonus	+	-
Myoclonus	+	-
Diarrhea	+	-

Radmard 2025

Mechanism	Class	Drugs implicated in serotonin syndrome
Inhibit serotonin synthesis	Dietary supplement	L-Tryptophan
Inhibit serotonin metabolism	Anxiolytics	Buspirone
	Herbal supplements	St. John's Wort (*Hypericum perforatum)*
	Monoamine oxidase inhibitors	Clorgyline, furazolidone, iproniazid, isocarboxazid, linezolid, methylene blue, moclobemide, pargyline, phenelzine, procarbazine, rasagiline, safinamide, selegiline, Syrian rue, tedizolid, and tranylcypromine
Increase serotonin release	Amphetamines + derivatives	Dexfenfluramine, fenfluramine, and phentermine
	Antidepressants (atypical)	Mirtazapine
	Cold remedies	Dextromethorphan
	Drugs of abuse	Cocaine, methylenedioxymethamphetamine (MDMA; Ecstasy)
	Opiates (not all)	Meperidine, oxycodone, and tramadol
	Parkinson disease Treatment/ amino acid	Levodopa

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Inhibit serotonin uptake	Amphetamines + derivatives	Dexfenfluramine, fenfluramine, and phentermine
	Antidepressants (atypical)	Bupropion, nefazodone, and trazodone
	Antiemetics	Granisetron and ondansetron
	Antihistamines	Chlorpheniramine
	Anxiolytics	Buspirone
	Cold remedies	Dextromethorphan
	Drugs of abuse	Cocaine, methylenedioxymethamphetamine (MDMA; Ecstasy)
	Herbal supplements	St. John's Wort (*Hypericum perforatum)*
	Opiates (not all)	Fentanyl, levomethorphan, levorphanol, meperidine, methadone, pentazocine, pethidine, propoxyphene, tapentadol, and tramadol
	Selective norepinephrine Reuptake inhibitors (SNRIs)	Desvenlafaxine, duloxetine, milnacipran, sibutramine, and venlafaxine
	Selective serotonin reuptake inhibitors (SSRIs)	Citalopram, escitalopram, fluoxetine, fluvoxamine, paroxetine, and sertraline
	Tricyclic antidepressants (TCAs)	Amitriptyline, amoxapine, clomipramine, desipramine, doxepin, imipramine, maprotiline, nortriptyline, protriptyline, and trimipramine

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Serotonin receptor agonists	Antidepressants (atypical)	Mirtazapine and trazodone
	Antiemetic/prokinetic agents	Metoclopramide
	Anxiolytics	Buspirone
	Drugs of abuse	Lysergic acid diethylamide (LSD)
	Ergot derivatives	Dihydroergotamine, ergotamine, and methylergonovine
	Mood stabilizers	Lithium
	Opiates (not all)	Fentanyl and meperidine
	Triptans	Almotriptan, eletriptan, frovatriptan, naratriptan, rizatriptan, sumatriptan, and zolmitriptan

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Mechanism	Class	Drugs implicated in serotonin syndrome
Serotonin receptor (5-HT2A) antagonists	Second-generation antipsychotics	Aripiprazole, clozapine, olanzapine, quetiapine, and risperidone
CYP450 microsomal oxidase interactions	CYP2D6	*Inhibitors*: fluoxetine and sertraline
		Substrates: Dextromethorphan, oxycodone, phentermine, risperidone, and tramadol
	CYP3A4	*Inhibitors*: ciprofloxacin and ritonavir
		Substrates: methadone, oxycodone, and venlafaxine
	CYP2C19	*Inhibitors*: fluconazole
		Substrates: citalopram
Modulators	Antidepressants (atypical)	Bupropion
	Parkinson's disease treatments	Amantadine, and levodopa

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Common situations 1

- Infection
  - Linezolid + MAO inhibitor or SSRI/SNRI
- Pain
  - Opioids + MAO inhibitor or SSRI/SNRI
- Substance use
  - MDMA or dextromethorphan + SSRI/SNRI

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Common situations 2

- HIV disease
  - SSRI especially fluoxetine + PI or NNRTI
- Cough, children
  - Dextromethorphan + SSRI/MAOi
- Surgery – methylene blue

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Common situations 3

- Herbal remedies
  - St. John's Wort *(Hypericum perforatum)*
  - Black seed oil
  - Ginseng
  - Brewer's yeast
  - Yohimbine

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Table 8.4 CYP-450 interactions of some serotonergic agents [52]

CYP enzyme	Substrates	Inducers	Inhibitors
1A2	Duloxetine	–	Fluvoxamine
2B6	Bupropion, methadone, selegiline, and sertraline	–	Paroxetine and sertraline
2C9		–	Fluvoxamine
2D6	Clozapine, codeine, desipramine, dextromethorphan, fluoxetine, haloperidol, hydrocodone, oxycodone, paroxetine, risperidone, selegiline, tricyclic antidepressants, and venlafaxine	–	Bupropion, fluoxetine, and paroxetine
3A4	Alfentanil, buspirone, cocaine, and methadone	St. John's Wort	–

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Low risk

- Parkinson's disease
  - MAO-B inhibitor use – low risk

- Migraine
  - Triptans/ergots + SSRIs/SNRIs

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Management

- Discontinue offending agent
- Fluids
- Benzodiazepines
- Cyproheptadine 12 mg stat then 4-8 mg q6h

Gandhi et al 2020

Table 8.6 Management of serotonin syndrome

Prompt recognition
Supportive care to control agitation, hyperthermia, and autonomic dysfunction
Discontinuation of all serotonergic agents
Intensive care unit monitoring, if needed
External cooling
Muscular paralysis with neuromuscular blocking agents
Mechanical ventilation
Sedation and muscle relaxation with intravenous benzodiazepine
Nonspecific serotonin receptor blockers, such as cyproheptadine, chlorpromazine, and methysergide
Electroconvulsive therapy may be considered

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Malignant catatonia

What is it?

- Catatonia = complex psychomotor syndrome
  - Lack of movement – inc. catalepsy/waxy flexibility
  - Lack of communication
  - >/=3 of: stupor, waxy flexibility, catalepsy, mutism, posturing, negativism, stereotypes, mannerisms, grimacing, agitation, echopraxia, and echolalia
  - +/- agitation, confusion, restlessness

- Akinetic, agitated, or catatonic

Edinoff et al 2021

Malignant catatonia

Catatonia

+

Rigidity

+

Hyperthermia

+

Autonomic instability

Apetauerova et al 2021

Table 7.1 Clinical features of malignant catatonia

Signs and symptoms
Hyperthermia
Catatonic excitement and/or stupor
Other catatonic features (e.g., mutism, negativism, catalepsy, posturing, echolalia, echopraxia, and staring)
Muscular rigidity (variable)
Altered consciousness
Autonomic instability
Profuse diaphoresis
Tachycardia
Labile or elevated blood pressure
Tachypnea and cyanosis (variable)
Positive laboratory findings
Most consistent-CPK elevation, leukocytosis
Less consistent low serum iron levels, elevated serum creatinine, hyponatremia, hypernatremia, dehydration, lymphocytic pleocytosis, EEG-generalized slowing, epileptic activity, delta brush, MRI hyperintensities in cortical and subcortical brain regions
Outgrowth of diverse neuromedical, drug-induced, and psychiatric conditions

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Associations

- Bipolar disorder
- Major depressive disorder
- Schizophrenia
- Autoimmune encephalitis - anti NMDA antibody associated
- Paraneoplastic limbic encephalitis
- Viral encephalitis

Psychiatric and neurodevelopmental disorders
Schizophrenia—9 cases
Mania—6 cases
Major depressive disorder—5 cases
Brief psychotic disorder—2 cases
Schizoaffective disorder—1 case
Autistic spectrum disorder—2 cases
Autoimmune disorders
NMDA receptor encephalitis —40 cases
GABA-A receptor encephalitis—2 cases
LGI1 receptor encephalitis—2 cases
Progressive encephalomyelitis with rigidity and myoclonus—1 case
Systemic lupus erythematosus—4 cases
Hashimoto's encephalopathy—2
Sjogren's syndrome—2 cases

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Infectious disorders
Viral encephalitis
Dengue virus —1 case
Herpes simplex virus type 1—1 case
Unspecified—3 cases
Encephalitis lethargica —2 cases
Mycoplasma pneumonia encephalitis —1 case
Neurobrucellosis encephalitis —1 case
Sepsis-associated encephalopathy—1 case

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Toxic and drug-related disorders
Clozapine withdrawal—3 cases
Amantadine withdrawal—1 case
Lorazepam withdrawal—1 case
Dexamethasone induced—1 case
Lithium toxicity—1 case
Cocaine-induced leukoencephalopathy—1 cases
Bath salts abuse—1 case
Methadone overdose—1 case
Disulfiram overdose—1 case
3-methoxyphencyclidine intoxication—1 case
Synthetic cannabinoid intoxication—1 case
Cyclosporine A-related neurotoxicity—1 case

Frucht S (Ed.) 2022. Movement Disorder Emergencies

	Participants (N)	Cases of catatonia (n [%])
Dalmau et al (2008)[72]	100	88 (88%)
Tsutsui et al (2012)[70]	3	2 (67%)
DeSena et al (2014)[73]*	8	5 (63%)
Kruse et al (2015)[74]	12	9 (75%)
Duan et al (2016)[75]	28	19 (68%)
Granata et al (2018)[76]*	18	8 (44%)
Herken and Prüss (2017)[77]†	53	10 (19%)
Total	222	141 (64%)

NMDAR=N-methyl-D-aspartate receptor. *All paediatric cases. †Relied on retrospective analysis of charts, so probably underestimated prevalence of catatonia.

Table 5: **Prevalence of catatonia (as identified by authors) in case series of NMDAR encephalitis**

Rogers et al 2019

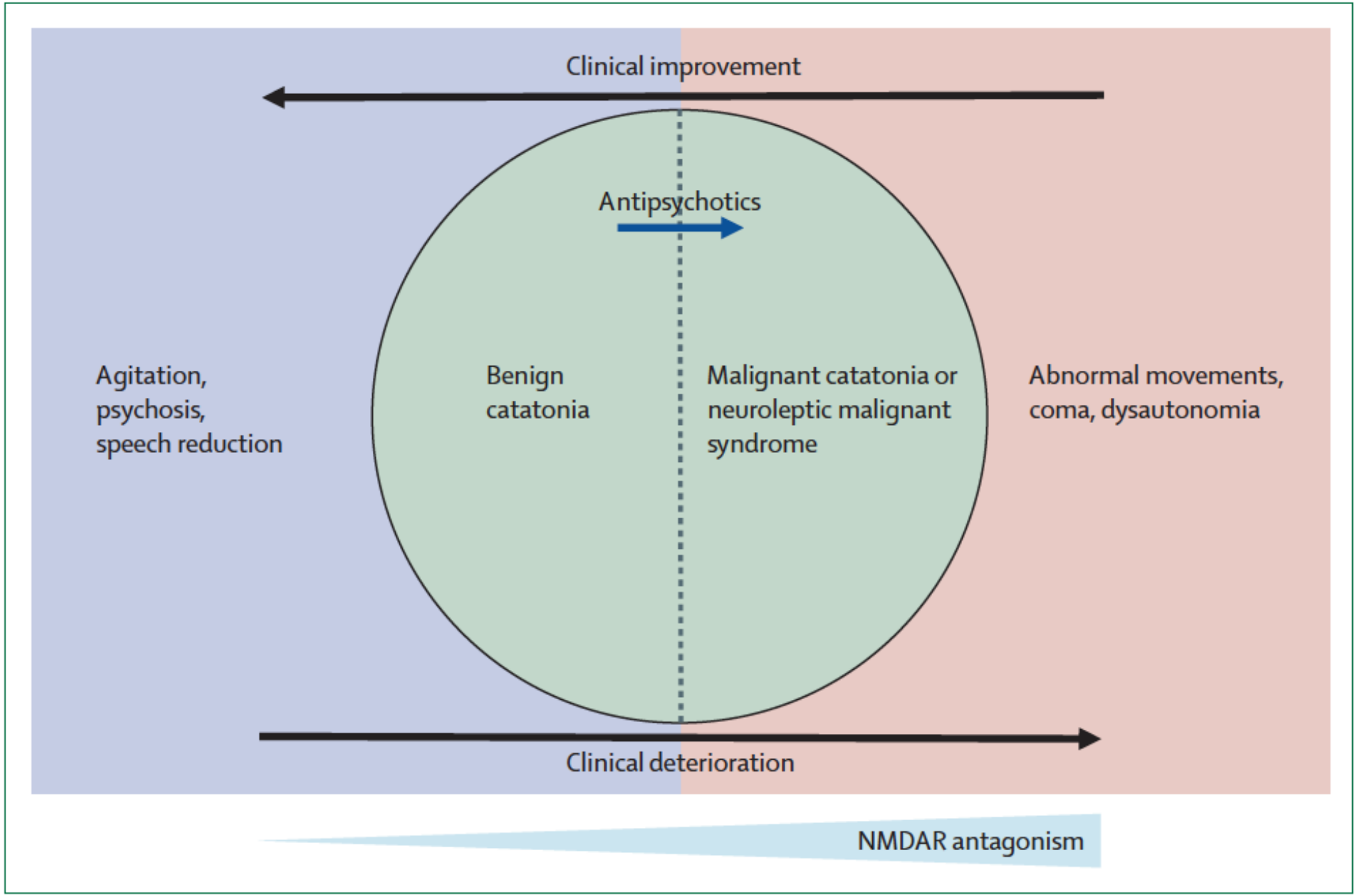
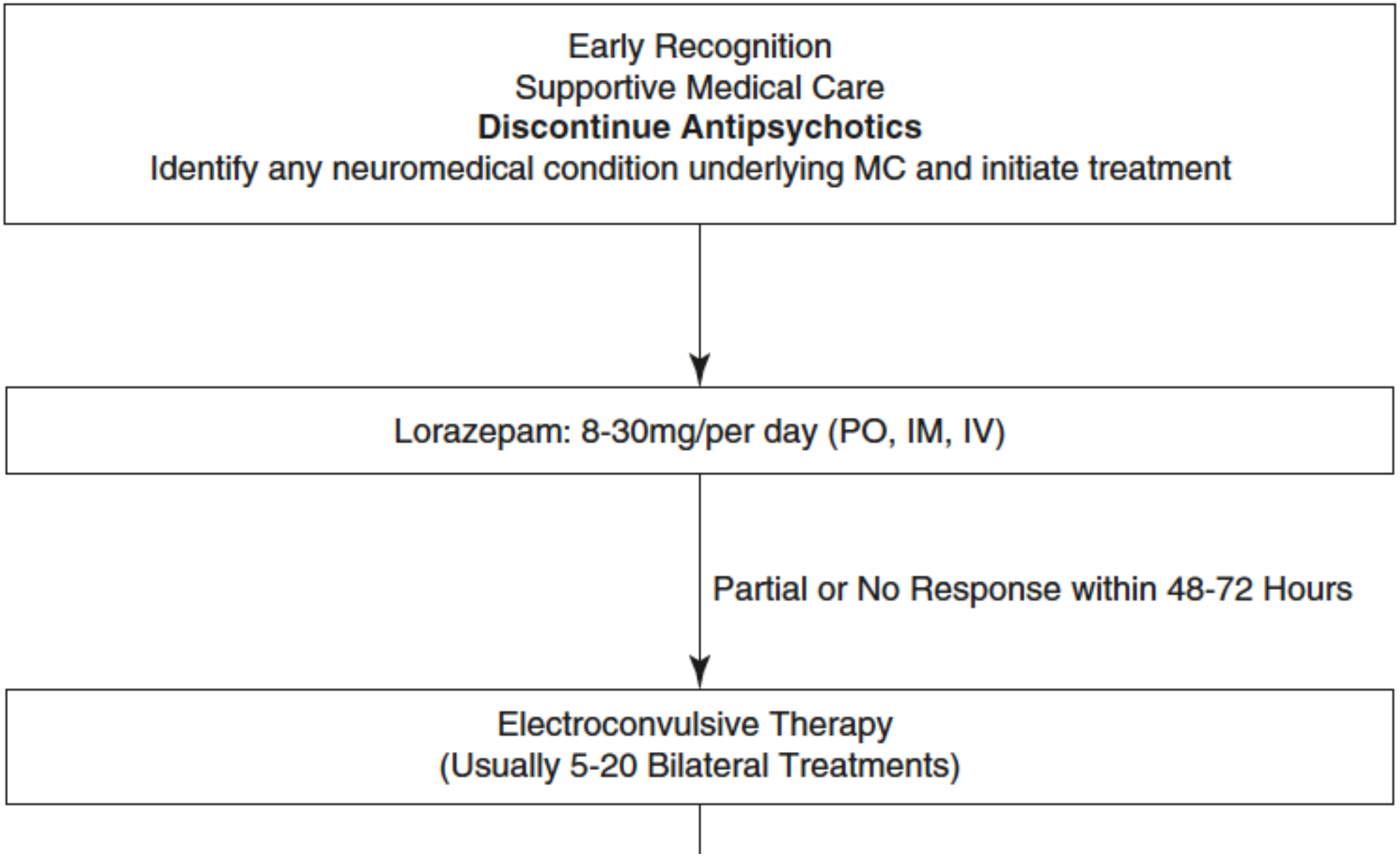


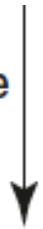
Figure: A model for glutamatergic hypofunction in catatonia

Rogers et al 2019



Frucht S (Ed.) 2022. Movement Disorder Emergencies

Partial or No Response



Treatments Supported by Anecdotal Evidence Derived from Case Reports

If stopped, restart lorazepam: 8-30mg (PO, IM, IV)
If significant rigidity and severe hyperthermia present add dantrolene: 1-2.5 mg/kg IV every 6 hours for 48 hours. If no improvement, switch to amantadine: 100mg PO or by nasogastric tube [NG] tube every 8 hours X 72 to 96 hours.
If partial or no improvement, switch to bromocriptine 2.5-5mg PO or by NG tube every 8 hours X 72 to 96 hours. If partial or no improvement, add memantine: 10mg per day PO or NG tube x 1 day, then 10mg BID x 5 days.
If no improvement, add propofol 25mcg/kg/min IV to 150mg mcg /kg/ min/IV
Add propofol: 25mcg/kg/min IV to 150 mcg/kg/min IV

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Acute dystonic reaction

Table 2.4 Description, etiologies, and treatments of oculogyric crisis

Characteristics	Etiologies	Treatment
Sustained, upward deviation of eyes lasting seconds to hours Ranges from mild and brief to severe, prolonged, and painful May have neck flexion, jaw opening, blepharospasm, and autonomic symptoms Psychiatric symptoms may be present	Drug induced: Typical and atypical neuroleptics Antiemetics Antidepressants Anticonvulsants Disorders of dopamine metabolism Hereditary and sporadic movement disorders Brain lesions: Dorsal midbrain Substantia nigra Basal ganglia Posterior third ventricle	Remove or reduce the dose of the offending agent Anticholinergics: Benztropine Biperiden Antihistamines: Diphenhydramine L-dopa may be beneficial in patients with parkinsonism

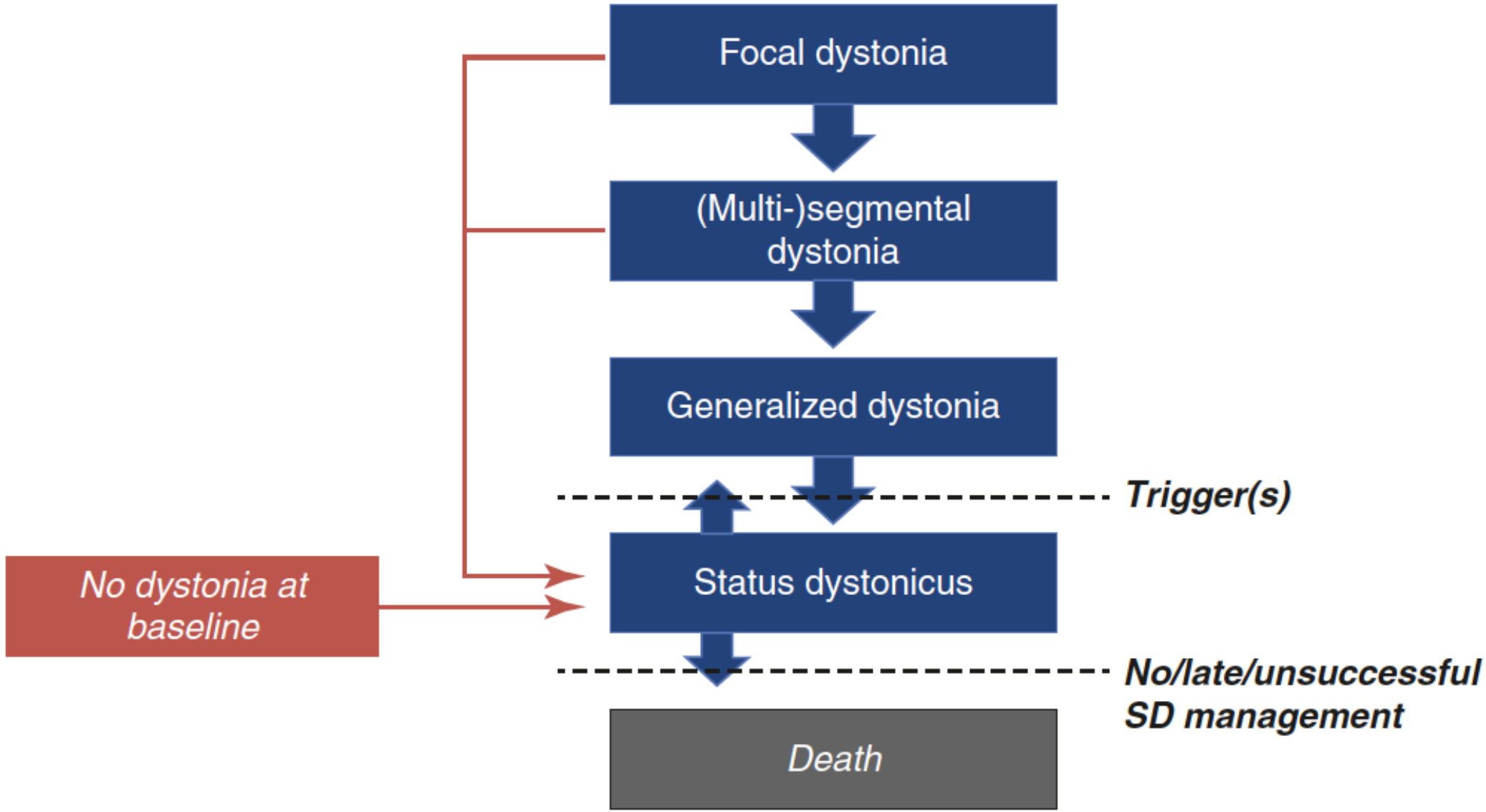
Frucht S (Ed.) 2022. Movement Disorder Emergencies

Status Dystonicus/Dystonic Storm

Definition

A life-threatening dystonic event associated with the development of one or more of the following (1) bulbar weakness, (2) respiratory failure, (3) metabolic derangements, and (4) exhaustion and pain + dystonia (Manji et al 1998)

A movement disorder emergency characterized by severe episodes of generalized or focal hyperkinetic movements that have necessitated urgent hospital admission because of the direct life-threatening complication(s) of these movements (Ruiz-Lopez and Fasano 2017)



Frucht S (Ed.) 2022. Movement Disorder Emergencies

Table 10.1 Focal/segmental dystonia as emergency

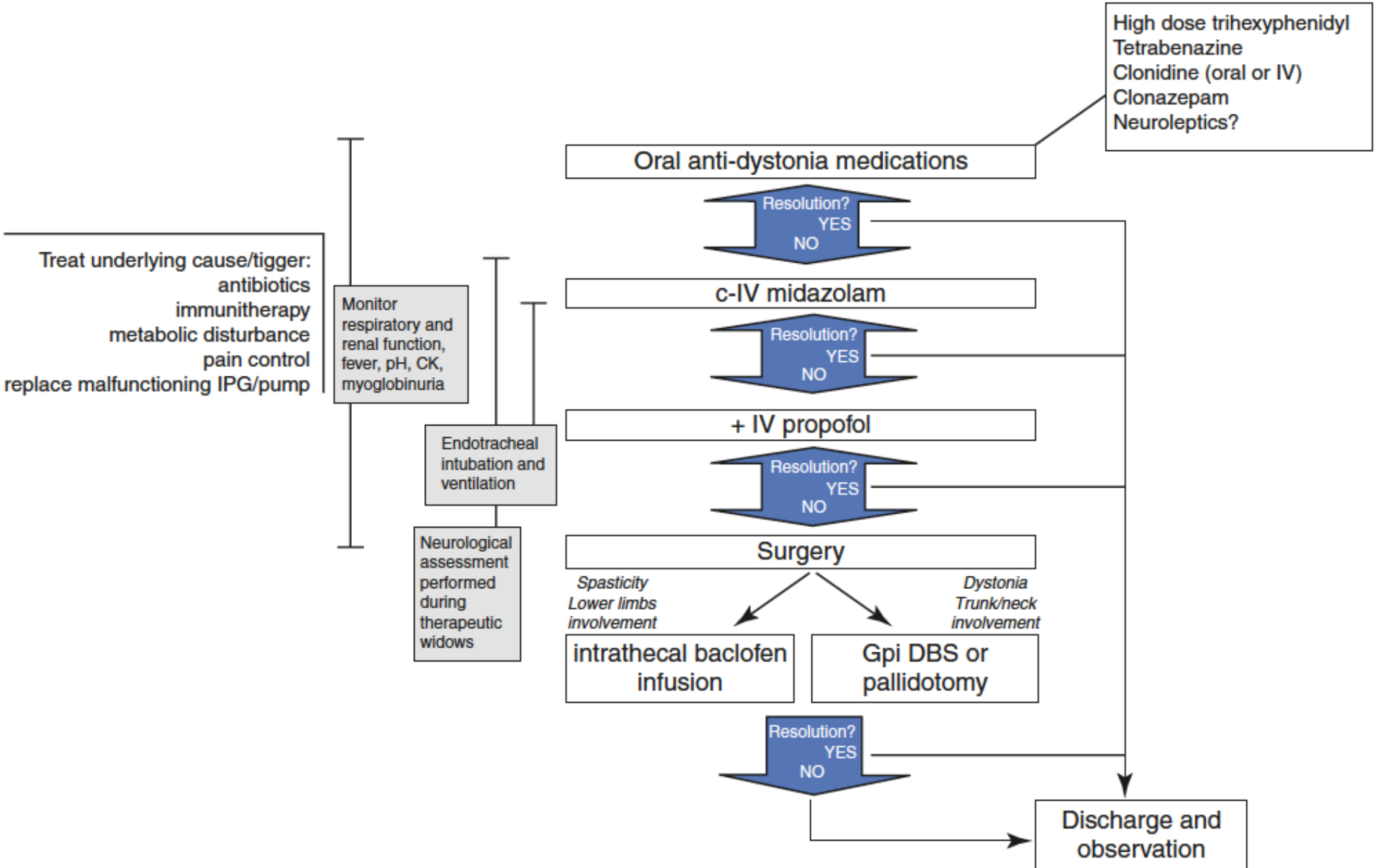
Pathophysiology	Causes	Phenomenology
Primary disorders of CNS	Paroxysmal dystonia	Usually involving a lower limb
Lesions	Basal ganglia stroke (especially putamen)	Usually involving trunk
Drug induced	D2 receptors antagonists (neuroleptic and antiemetics)	Usually involving trunk (e.g., Pisa syndrome) or lower face (e.g., oromandibular dystonia)
Degenerative	Multiple system atrophy	Adductor laryngeal dystonia
Psychiatric	Functional dystonia	Usually involving trunk (e.g., camptocormia) or upper limb
Mimickers	Atlantoaxial subluxation, posterior fossa and cervical cord lesions, retropharyngeal abscess, immune reactions.	Usually involving neck, laryngeal muscles (i.e., stridor), or upper limb (e.g., pseudo-dystonia due to somatosensory deficits).

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Table 10.5 The triggers of status dystonicus reported to date

Anesthesia
Autoimmune processes
Diet (protein loading in glutaric aciduria)
Hardware failure (DBS or ITB pump)
Infections
Medication changes Introduction of, e.g., penicillamine (WD), zinc (WD), dopamine-receptor blockers, clonazepam Withdrawal of, e.g., penicillamine (WD), dopamine depleting agents, baclofen, lithium
Metabolic disturbances
Puberty
Surgery (including DBS surgery)
Trauma (physical and psychological)

Frucht S (Ed.) 2022. Movement Disorder Emergencies



Frucht S (Ed.) 2022. Movement Disorder Emergencies

Post-anoxic myoclonus

Table 11.1 Characteristic diagnostic features in acute and chronic posthypoxic myoclonus

	Acute PHM	Chronic PHM
Time of onset	24–48 h	Upon regaining consciousness
Demographic	Adults	Adults
Type of myoclonus	Subcortical	Cortical and/or subcortical Negative myoclonus (mostly lower limbs)
Clinical features of myoclonus	Generalized, symmetric, proximal, stereotyped, stimulus sensitive Possible myoclonic status (myoclonus lasting continuously >30 min) Cranial nerves: caudocranial involvement Occasionally focal	Focal or multifocal, more distal, action and intentional myoclonus, stimulus sensitive Cranial nerves: craniocaudal involvement Occasionally focal
Additional neurological features	Severe general deterioration Cognitive impairment Frequent seizures	Ataxia Dysarthria Dysphagia Gait impairment Seizure is rare
EEG	Status epilepticus, burst suppression, diffuse slow waves and background, spike-wave activity, alpha coma	Spikes at the vertex or in the contralateral cortical motor area
Additional neurophysiology	EEG abnormalities not time-locked Normal SEPs Normal long latency reflex	± time-locked EEG abnormalities Giant SEPs Prolonged long latency reflex
IMAGING	Nonspecific	Nonspecific (brain MRI) PET scan: increased signal in VL nucleus (thalamus), pontine tegmentum, mesencephalon
PROGNOSIS	Poor (survival up to 10% cases)	Good (rare additional neurological sequela, possible improvement of myoclonus)

Frucht S (Ed.) 2022. Movement Disorder Emergencies

POST-HYPOXIC MYOCLONUS

ACUTE

FIRST LINE TREATMENTS

TTM (normothermia at 36°C for 24 hours, or therapeutic hypothermia at 32°C)

ANTIEPILEPIC MEDICATIONS:
Benzodiazepine (diazepam, clonazepam, lorazepam)
Phenytoin
Phenobarbital
Valproic acid

SECOND LINE TREATMENTS

Propofol
Isoflurane

CHRONIC

FIRST LINE TREATMENTS

Clonazepam (up to 15 mg/day)
Valproic Acid (up to 2000 mg/day)
Levetiracetam (up to 3000 mg/day)

> Start with single medication
> Combination of multiple drugs usually needed

SECOND LINE/ ADD ON TREATMENTS

L-5-HTP
Perampanel

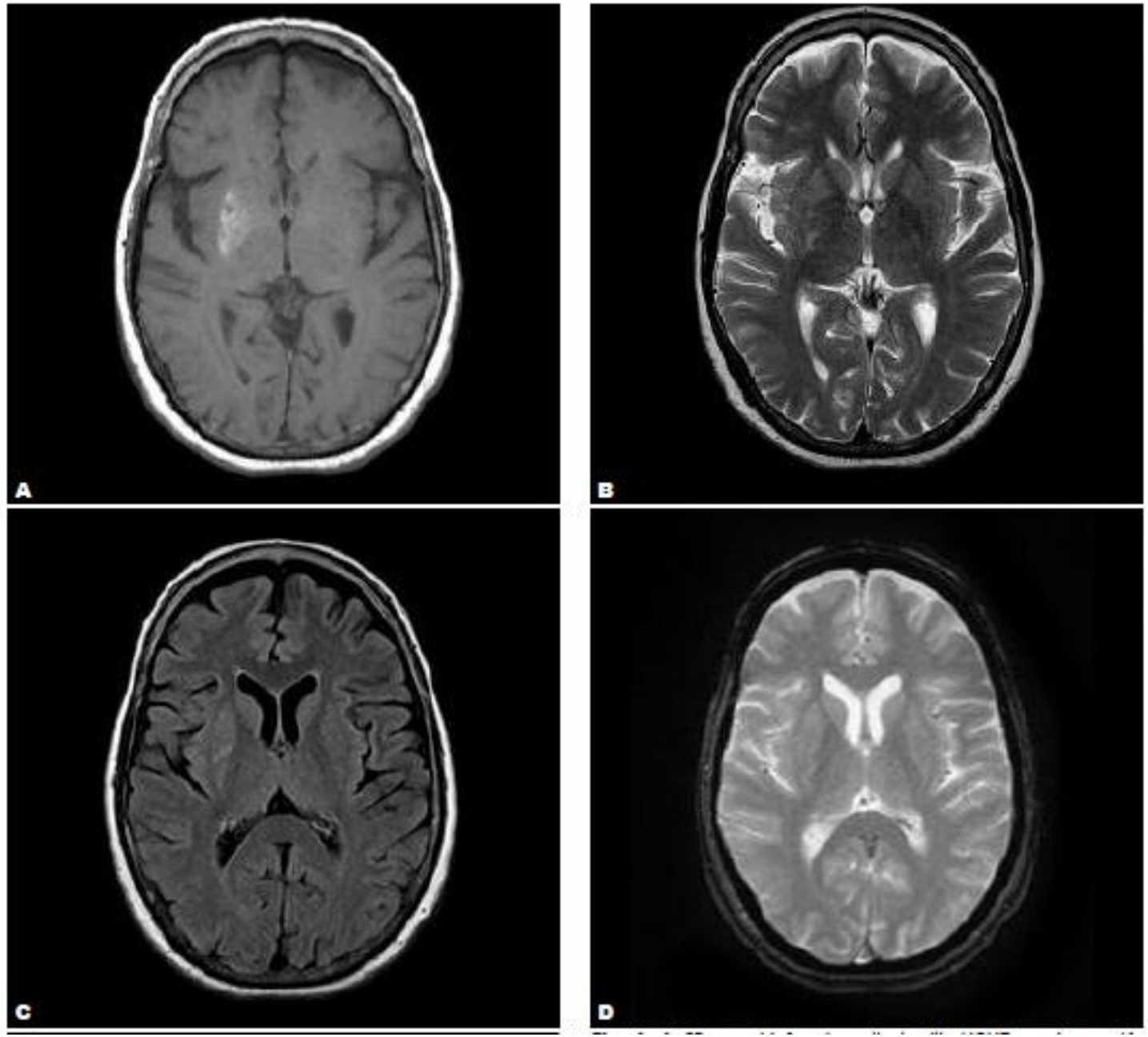
ISOLATED REPORTS:

Lacosamide
Zonisamide
Levodopa
Carbamazepine
Chlorpromazine
Paroxetine
Estrogen
Agomelatine
Cannabidiol
Intratecal baclofen

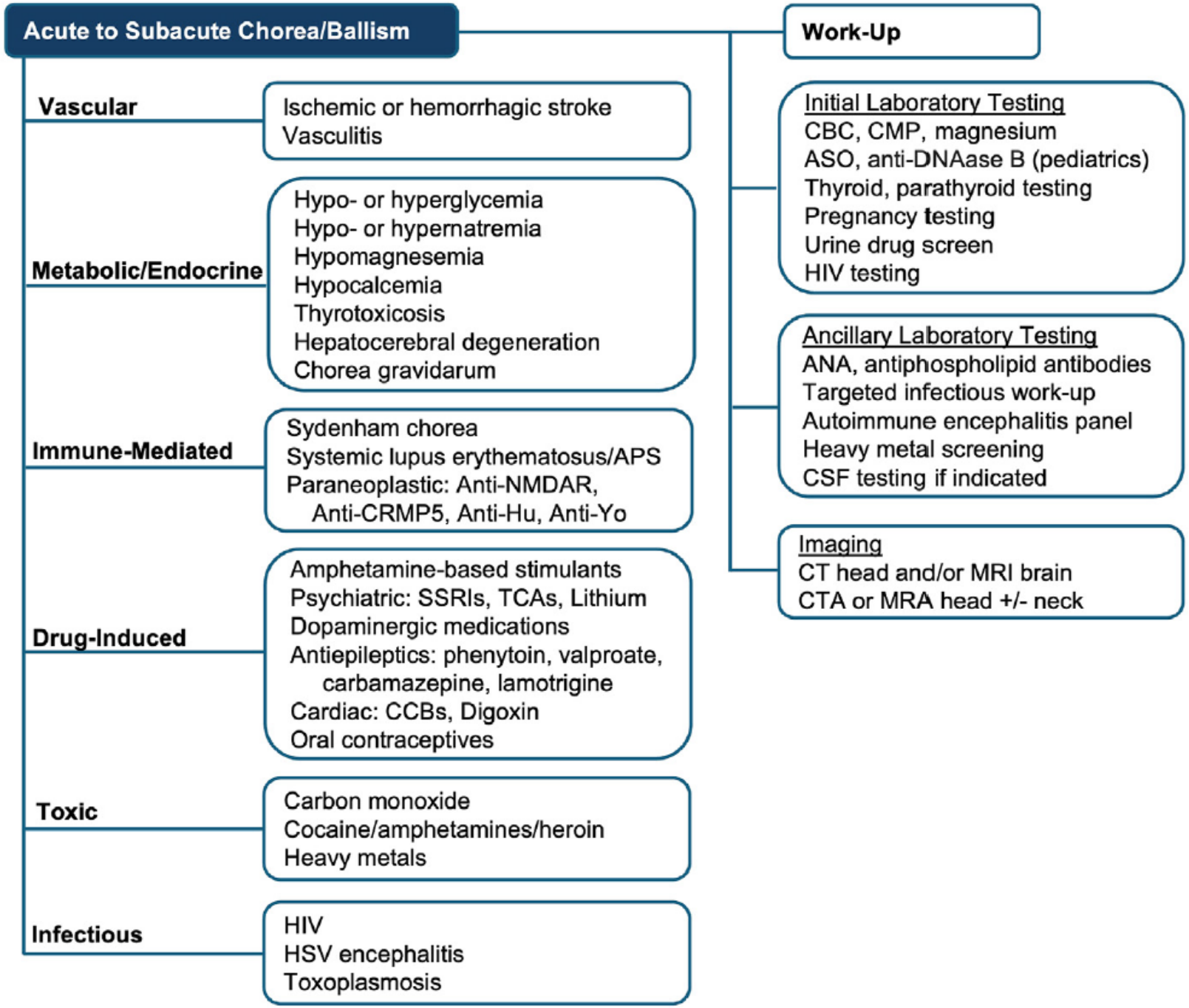
THIRD LINE TREATMENTS

Sodium Oxybate (add-on)
DBS (GPi)

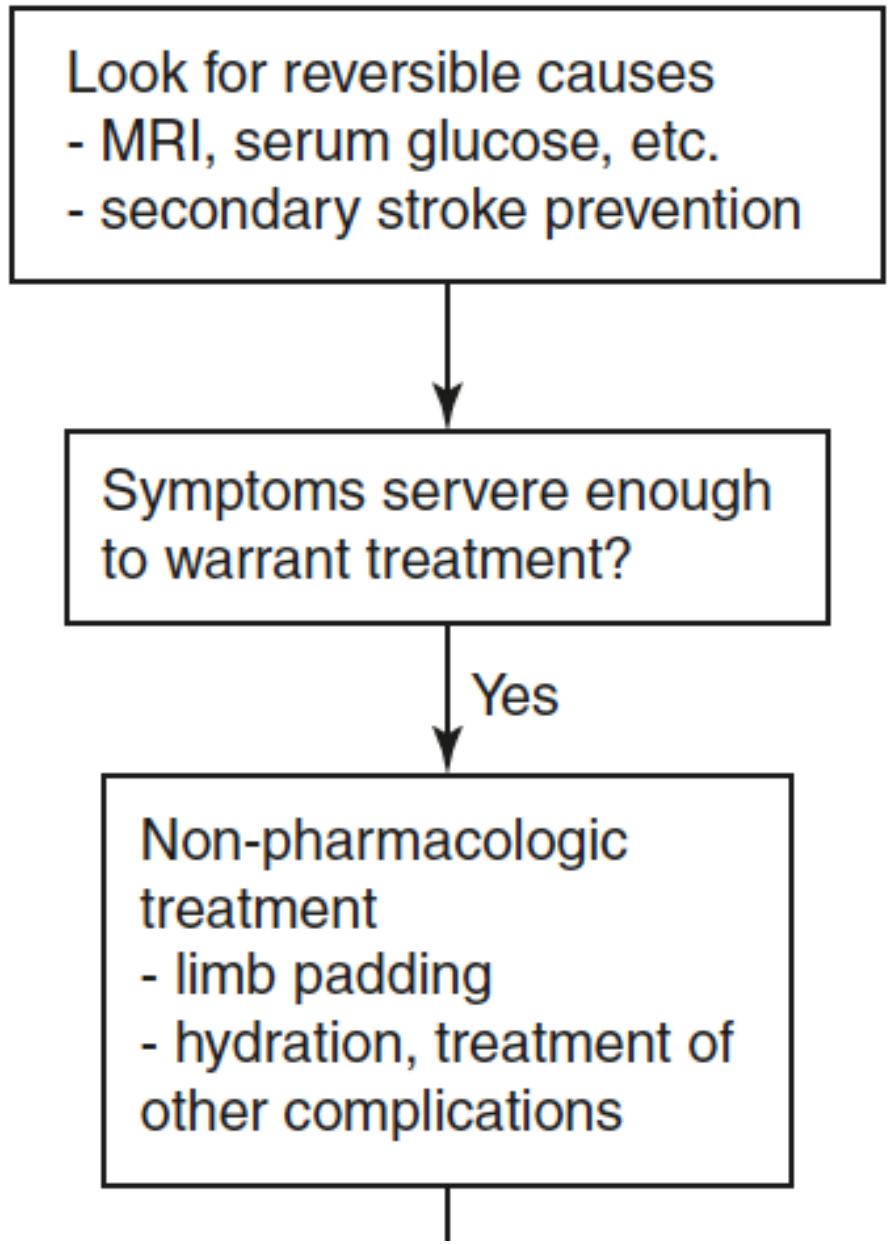
Hemiballism-hemichorea



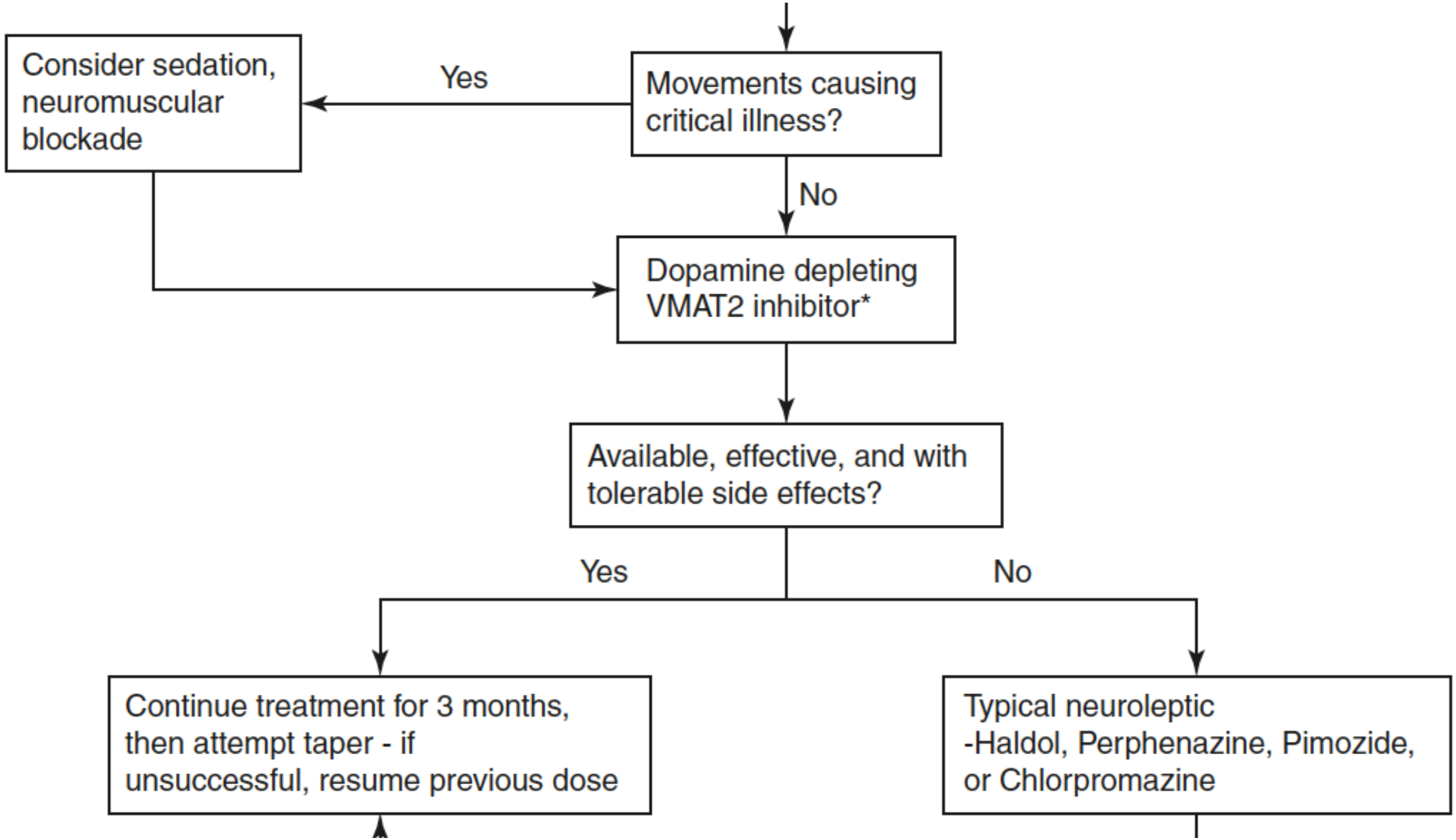
Mittal 2011



Radmard 2025



Frucht S (Ed.) 2022. Movement Disorder Emergencies



Frucht S (Ed.) 2022. Movement Disorder Emergencies

Tic storm

Internal
Fatigue
Hormone status
Level of perceived stress
External
Diet
Drugs
Infections

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Table 12.4 Selected drugs to treat tics

Drug	Usual adult starting dose	Usual maximum dose/day
Pimozide	1 mg at bedtime	10 mg
Haloperidol	0.25 mg at bedtime	20 mg
Aripiprazole	5 mg once daily	20 mg
Fluphenazine	0.5 mg at bedtime	5 mg
Risperidone	0.25 mg at bedtime	4 mg
Tetrabenazine	12.5 mg at bedtime	200 mg
Deutetrabenazine	6 mg once daily	48 mg
Valbenazine	40 mg once daily	80 mg
Clonidine	0.05 mg at bedtime	0.8 mg
Guanfacine	0.5 mg at bedtime	3 mg
Botulinum toxin	Varies with injected muscle	Varies with injected muscle

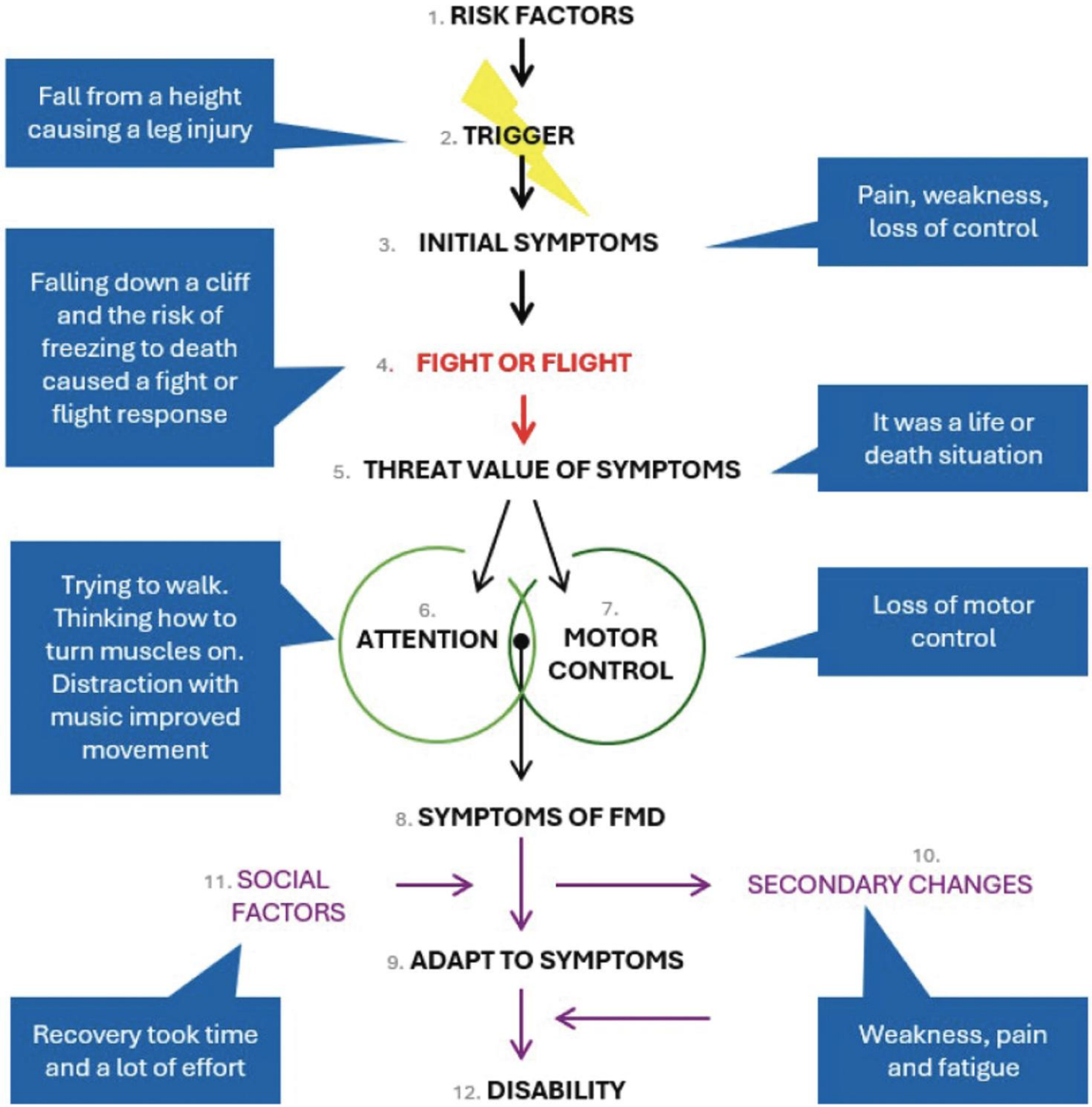
Frucht S (Ed.) 2022. Movement Disorder Emergencies

Functional movement disorder emergency

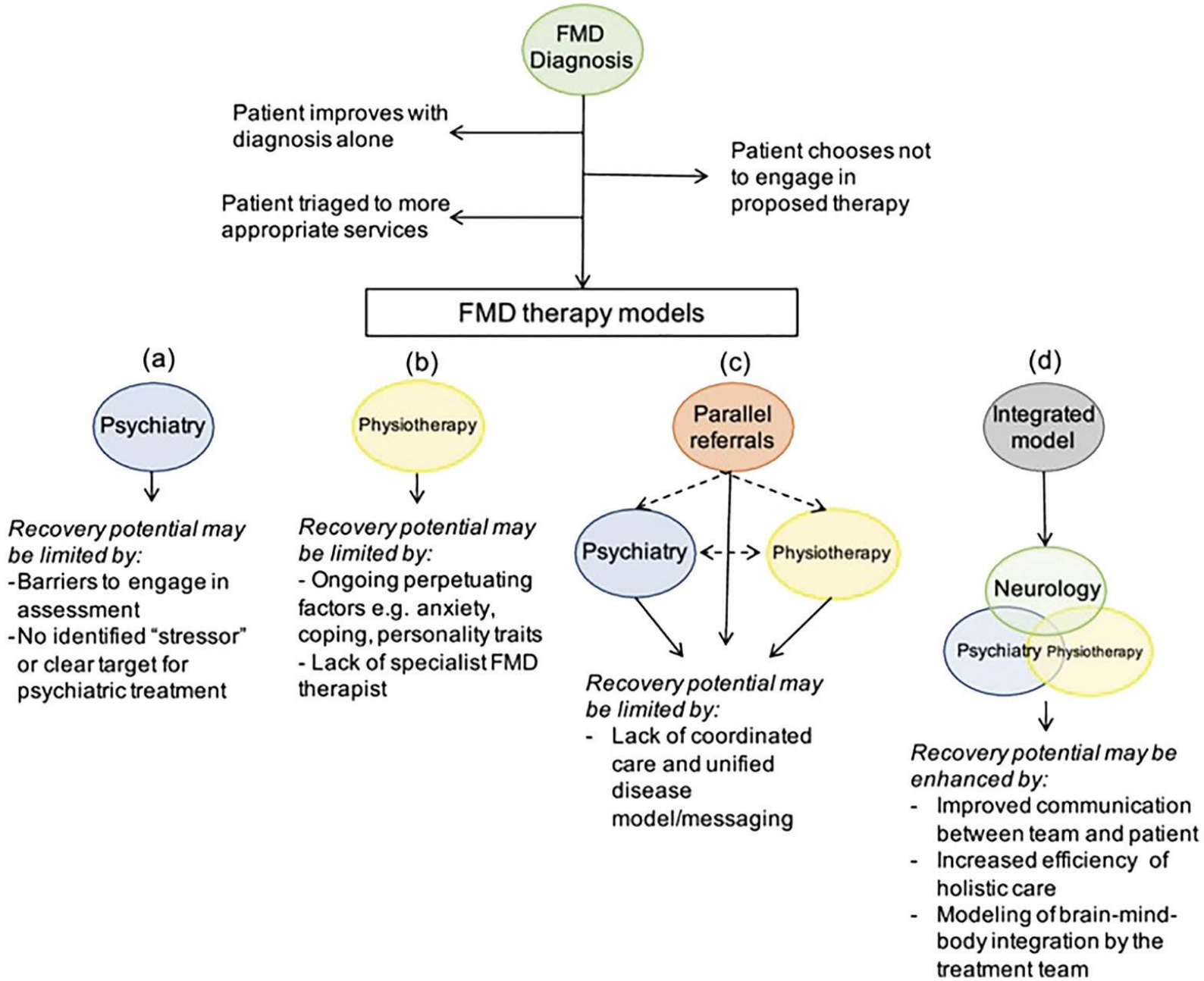
Table 7
Core diagnostic criteria of functional movement disorder[73]

Diagnostic Criteria	Description
Variability	Presence of inconsistent frequency, duration, or direction of the presenting abnormal movement
Distractibility	Severity of movement disorder lessens or resolves with distraction maneuvers, like serial sevens, saying the months of the year backwards, or performing other examinations maneuvers
Entrainment	Frequency of the abnormal movement syncs with the frequency of a repetitive task, like finger taps, hand opening/closing, or toe taps
Suggestibility	Direct mention of the abnormal movement or third person presence triggers the abnormal movement

Radmard 2025

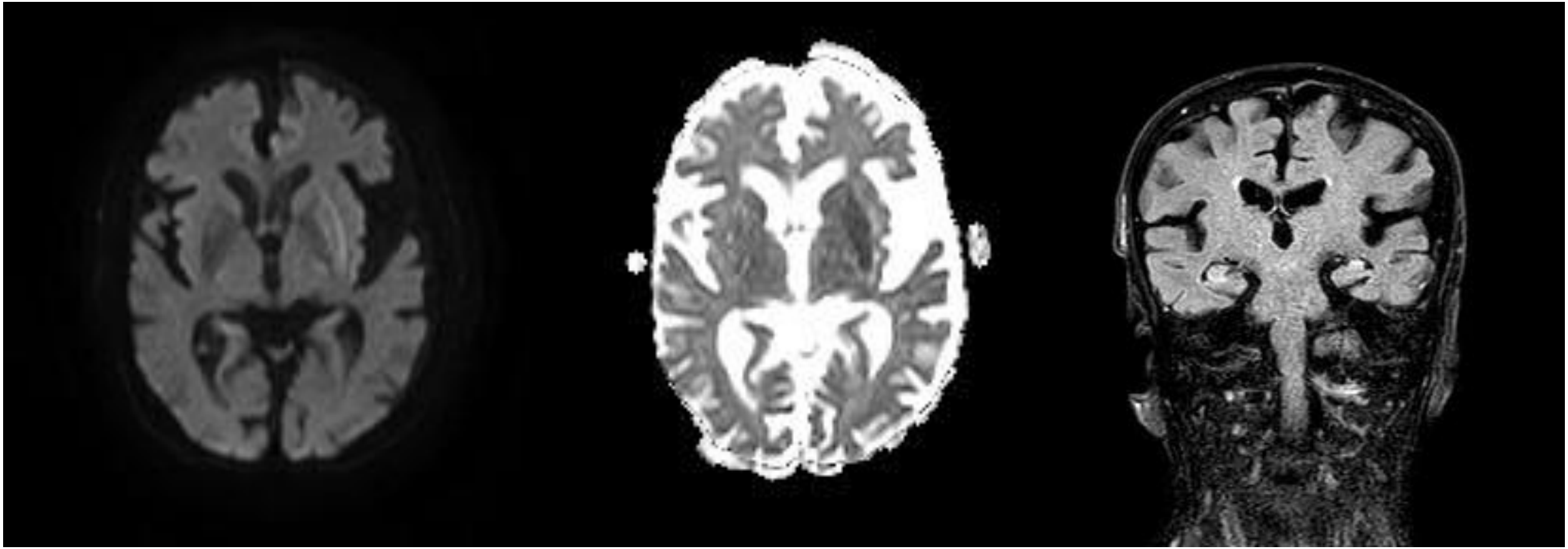


Stone and Carson 2025



Holden and Madera 2022

Autoimmune encephalitis



Ataxia	Chorea	Dystonia	Myoclonus	Parkinsonism	Stiff-person syndrome
Paraneoplastic cerebellar degeneration (*Yo, -Hu, –Tr, DNER, Ri*)	Autoimmune encephalitis (*NMDAR*, Neurexin3, LGI1, CASPR2, IgLON5)	Autoimmune encephalitis (*NMDAR*)	Opsoclonus myoclonus ataxia syndrome	Basal ganglia encephalitis (children)	Stiff person syndrome (*GAD65, Glycine receptor, Amphiphysin*)
Autoimmune ataxia (*GAD65*, CASPR2, GluR1)	Post-HSV autoimmune encephalitis (*NMDAR*, D2R)	Paraneoplastic dystonia (–Ri, IgLON5)	Autoimmune encephalitis (CASPR2, LGI1, glycine receptor, DPPX, IgLON5)	Paraneoplastic parkinsonism (*CRMP5, –Ma*)	Progressive encephalomyelitis and rigidity myoclonus (PERM) (*Glycine receptor, GAD65*, DPPX)
	Paraneoplastic chorea (*CRMP5*, Hu)			Autoimmune encephalitis (NMDAR, D2R, DPPX glycine receptor, GAD6S, LGI1, CASPR2, IgLON5)	

Frucht S (Ed.) 2022. Movement Disorder Emergencies

Box 1
Diagnostic criteria for possible autoimmune encephalitis[4]

1. Subacute onset (rapid progression of <3 months) of working memory deficits (short-term memory loss), altered mental status, or psychiatric symptoms
2. At least 1 of the following:
   - New focal central nervous system findings
   - Seizures not explained by a previously known seizure disorder
   - CSF pleocytosis (WBC>5 cells/mm^3)
   - MRI features suggestive of encephalitis
3. Reasonable exclusion of alternative causes

Abbreviation: WBC, white blood cell count.

Raibagkar and Ramineni 2021

Box 3
Diagnostic criteria for anti–*N*-methyl-D-aspartate receptor encephalitis[4]

Probable

1. Rapid onset (<3 months) of at least 4 of the 6 following major groups of symptoms:
   - Abnormal (psychiatric) behavior or cognitive dysfunction
   - Speech dysfunction (pressured speech, verbal reduction, mutism)
   - Seizures
   - Movement disorder, dyskinesias, or rigidity/abnormal postures
   - Decreased level of consciousness
   - Autonomic dysfunction or central hypoventilation
2. At least 1 of the following:
   - Abnormal EEG (focal or diffuse slow or disorganized activity, epileptic activity, or extreme delta brush)
   - CSF with pleocytosis or oligoclonal bands
3. Reasonable exclusion of other disorders

   Diagnosis can be made in the presence of 3 of the above group symptoms accompanied by a systemic teratoma

Definite

1. Diagnosis can be made in presence of 1 or more of the 6 major group of symptoms and immunoglobulin (Ig) G anti-GluN1 antibodies, after reasonable exclusion of other disorders

Raibagkar and Ramineni 2021

FIRST LINE THERAPIES

IV Methylprednisolone 1gm x 3-5d

(caution if concern for infection or CNS lymphoma)

AND

Intravenous Immunoglobulin (IVIG)

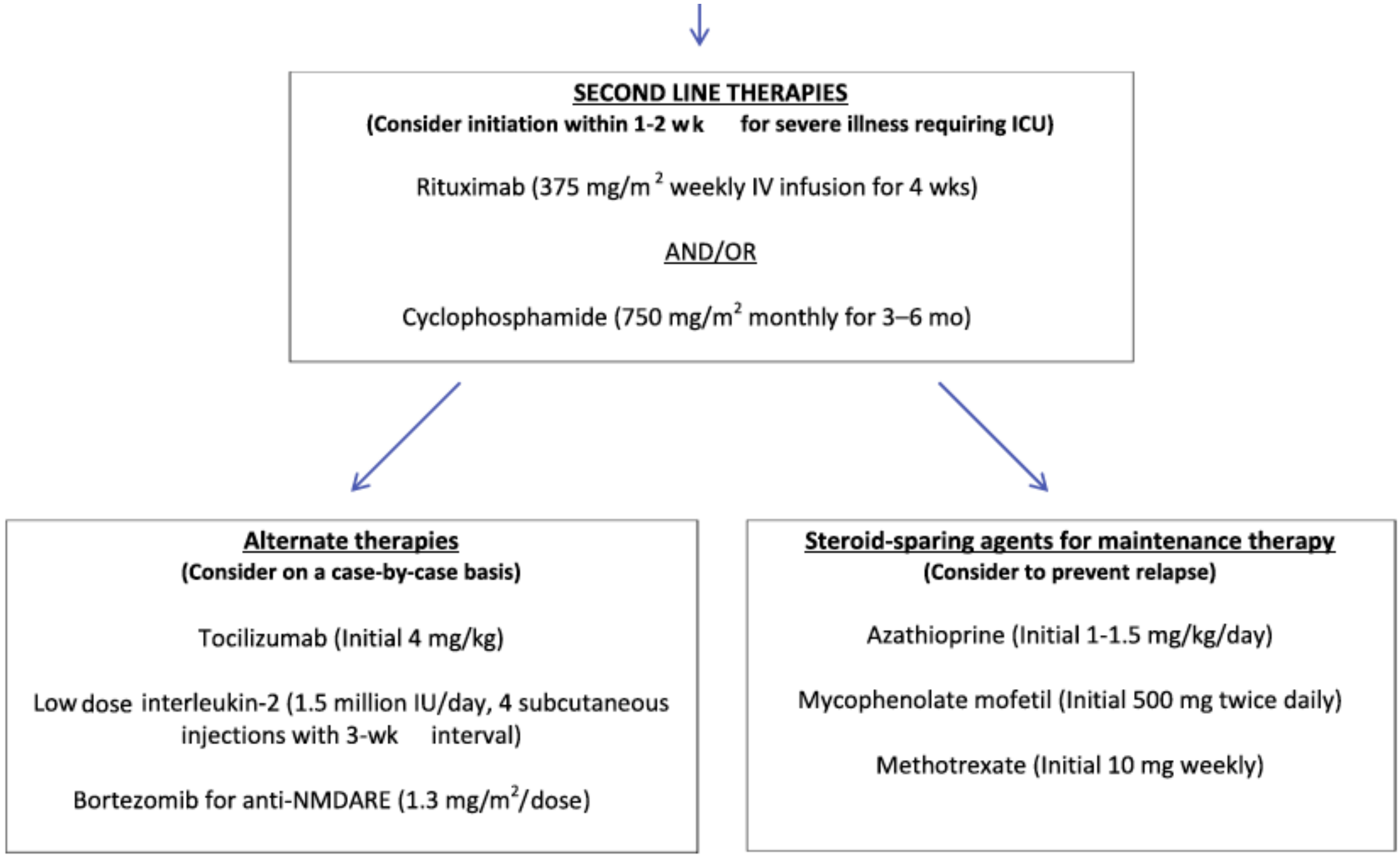
AND/OR

Plasma Exchange (PLEX)

AND

Early removal of tumor if present

Raibagkar and Ramineni 2021



Raibagkar and Ramineni 2021

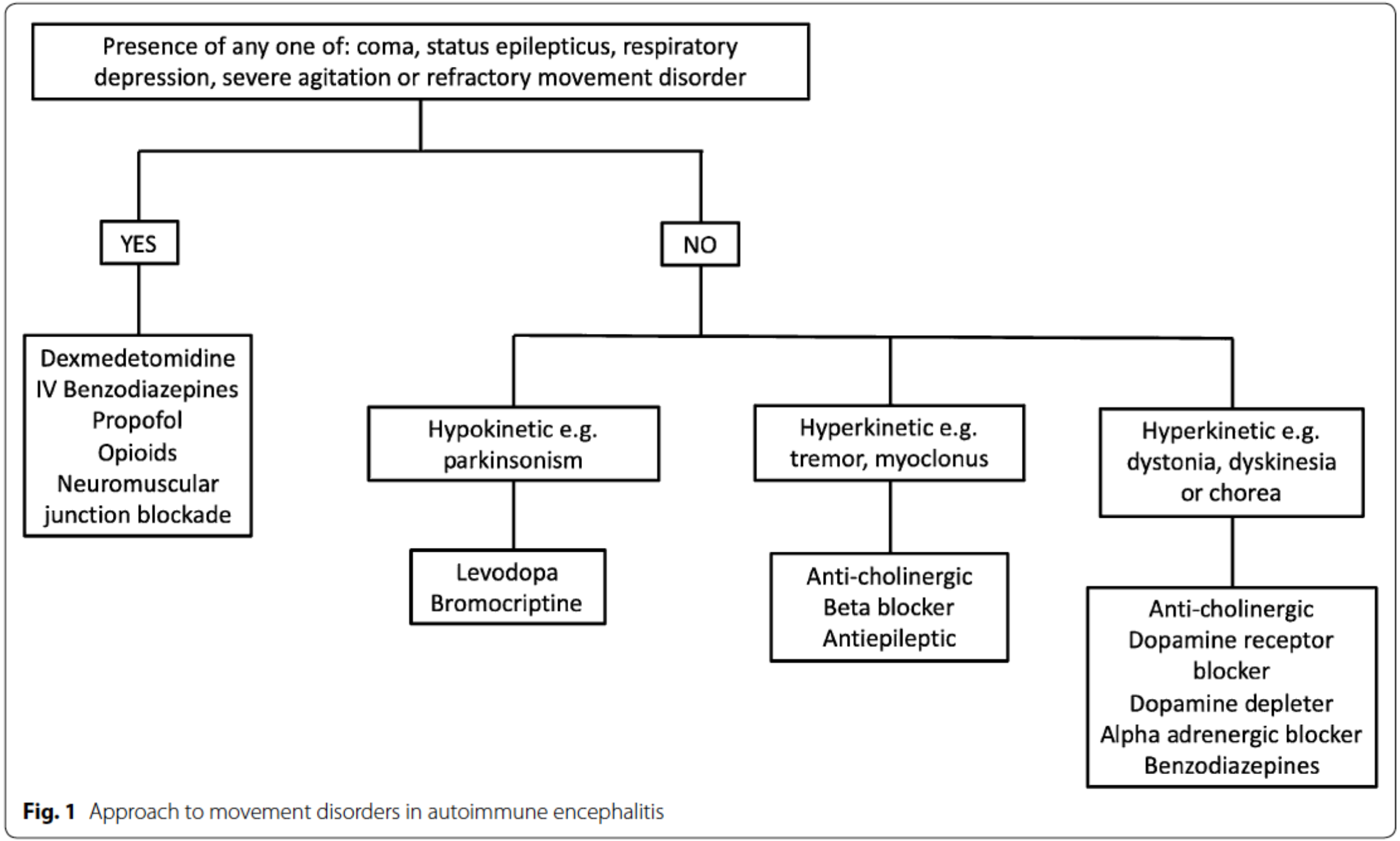


Fig. 1 Approach to movement disorders in autoimmune encephalitis

Ali and Wijdicks 2019

https://www.artofemergencymedicine.com/volume-15