CEREBELLAR ATAXIA CHEAT SHEET

Cerebellar Ataxia

A Vestibular Physician's quick reference — recognition, localisation, work-up and management.

► Why cerebellar ataxia matters

The most commonly missed and most time-critical cause of imbalance reaching a balance clinic. Cerebellar ataxia is a syndrome, not a diagnosis: confirm the cerebellar localisation, then find the cause along the hereditary / sporadic-degenerative / acquired / immune axis. An acute cerebellar stroke can present exactly like vestibular neuritis — the bedside discrimination is life-saving.

Recognising the cerebellar pattern

- Wide-based gait ataxia, impaired tandem walking, truncal titubation.
- Appendicular dysmetria, dysdiadochokinesis, intention tremor.
- Scanning dysarthria; gaze-evoked or downbeat nystagmus.
- Cerebellar cognitive affective syndrome (CCAS) — executive, visuospatial and affective change. Screen for it; the cerebellum is not purely motor.
- Multisystem clues: neuropathy + pyramidal signs (Friedreich ataxia), slow saccades (SCA2), autonomic failure (MSA).

Localisation — zone to dominant signs

Functional zone	Dominant signs
Vestibulocerebellum (flocculonodular lobe)	Nystagmus, vertigo, impaired smooth pursuit and VOR
Spinocerebellum (vermis / paravermis)	Wide-based gait ataxia, truncal titubation
Cerebrocerebellum (lateral hemispheres)	Limb dysmetria, intention tremor, dysarthria, CCAS

Onset tempo — the first diagnostic filter

Tempo	Likely causes
Acute (hours–days)	Stroke / haemorrhage, intoxication, Wernicke, post-infectious cerebellitis
Subacute (weeks–months)	Immune / paraneoplastic, infective, toxic-metabolic, prion disease
Chronic (months–years)	Hereditary SCA / FRDA, MSA-C, sporadic degenerative, CANVAS

Classification — the genetic groups

Group	Flagship genotypes	Key features
Autosomal dominant (SCAs)	SCA3 (commonest), SCA6 (pure/slow), SCA2 (slow saccades), SCA7 (retinopathy)	Adult-onset CAG repeat expansions
Autosomal recessive	Friedreich ataxia (FXN), ataxia-telangiectasia	Childhood/teen onset; multisystem; several treatable
X-linked	FXTAS (FMR1 premutation)	Late-onset tremor-ataxia, males predominate
Mitochondrial	POLG, MELAS, MERRF, NARP	Ataxia + epilepsy, neuropathy, ophthalmoplegia
New & testable	CANVAS (RFC1), SCA27B (FGF14)	Common late-onset; shrink the 'idiopathic' group

Investigations — tiered

Tier	Tests	When
First-line	MRI brain (± spine), B12, TFT, vitamin E, glucose; toxin + thiamine if acute	All patients
Second-line	Autoimmune/paraneoplastic antibodies, anti-GAD/TG6, infection serology, EMG/NCS, LP	Subacute or immune suspicion
Third-line	Repeat panels (SCA, FXN, RFC1), whole-exome sequencing, metabolic assays	Hereditary/young or undiagnosed

Pearl — *MRI is highest-yield: 'hot cross bun' + middle-cerebellar-peduncle change suggests MSA-C; pure cerebellar atrophy favours an SCA.*

Differential diagnosis

Pattern	Distinguishing features
Cerebellar ataxia	Vision-independent; dysarthria, dysmetria, gaze-evoked/downbeat nystagmus
Sensory ataxia	Romberg-positive, vision-dependent; no dysarthria or nystagmus; distal sensory loss
Vestibular ataxia	Vertigo; unidirectional, fixation-suppressed nystagmus; directional veer
Functional gait	Inconsistent, distractible; coordination normal on formal testing

► Red flags — stroke and urgent referral

Acute cerebellar ataxia with vertigo is a posterior-circulation stroke until proven otherwise. A normal horizontal head-impulse test with direction-changing nystagmus ± skew deviation (central HINTS) is more sensitive than early MRI — arrange urgent imaging and stroke pathways. Escalate urgently also for rapidly progressive ataxia, severe headache, depressed consciousness, or a new focal deficit.

Management

Tier	Intervention	Principles
Treat the cause	Immune (steroids / IVIG / PLEX), nutritional (B12, thiamine, vitamin E, copper), Wilson, CoQ10	Highest-value act; early treatment preserves cerebellar reserve
Rehabilitation	Physiotherapy (balance/gait), OT, speech, vestibular rehab	Cornerstone; start at diagnosis; durable SARA gains
Symptomatic drugs	4-AP / acetazolamide (EA2, nystagmus); riluzole; clonazepam, gabapentin, memantine	Individualise; review for benefit; taper if no gain
Aids & safety	Walking aids, home modification, falls and driving review	Avoid alcohol; multidisciplinary care

Pearl — *Always screen for the treatable ataxias — immune, nutritional, Wilson disease, CoQ10 deficiency — because delay converts a reversible disorder into a permanent one.*

Counselling and follow-up

- Prognosis is aetiology-dependent — from reversible (immune, nutritional) to relentless (MSA-C, ~6–10 yr survival).
- Genetic counselling for hereditary disease (dominant = 50% transmission); generally avoid testing asymptomatic minors.
- Record SARA at baseline and each review; re-interrogate 'idiopathic' diagnoses as RFC1 / FGF14 testing becomes available.
- Connect patients to rehabilitation, support groups, and trials (Ataxia Global Initiative).

Key refs — Harding 1983 · Klockgether 2010/2019 · Schmahmann 1998 · de Silva 2019 · Hadjivassiliou 2020 · Zesiewicz/AAN 2018 · Cortese 2019 (RFC1) · Strupp 2011/2017