Central Causes of Vestibular Dysfunction in Children:

Posterior Fossa Tumours, Stroke, and Demyelinating Disease

Vestibular Medicine in Children

Topic 6 of 15

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How to Use This Review

This literature review is part of the Vestibular Medicine in Children series published by the Australian Dizziness Clinics Education Hub. It is written for vestibular physicians, paediatricians, and emergency physicians who assess and manage children presenting with vestibular disorders.

The review is designed to be read as a deep-reference resource or used as a clinical desktop companion. It is supported by a clinical cheat sheet, short-form clinician videos, and audio episodes that cover the same material.

Callout Box Guide

☐ **Key Point:** Foundational concepts and summary statements that anchor the core clinical content of each section.

☐ **Clinical Insight:** Clinically relevant observations for direct application in assessment and management.

☐ **Clinical Pearl:** High-yield memorable clinical points — the take-home messages most likely to change practice.

☐ **Important:** Red flags, emergencies, and critical safety points requiring immediate action.

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VI. Clinical Features That Point to Central Pathology

Diagnosis	Management approach	Outcome
Posterior fossa tumour	Surgical resection ± adjuvant therapy; vestibular rehabilitation post-op	Depends on histology; VRT reduces balance morbidity
Posterior circulation stroke	Thrombolysis/thrombectomy if within window; anticoagulation for cardioembolic	Childhood stroke has better plasticity than adult
Demyelination (MS)	IV methylprednisolone 1 g/day × 3–5 days for acute relapse	Neurology manages; VRT for residual vestibular deficit
Cerebellar abscess	IV antibiotics + surgical drainage (mastoidectomy if otogenic)	Excellent if treated rapidly; ENT + neurosurgery
Chiari malformation	Surgical decompression if symptomatic; conservative if asymptomatic	Neurosurgery decision; annual MRI surveillance

VII. Neuroimaging in Paediatric Central Vestibular Disease

VIII. Vestibular Function Testing in Central Pathology

Sign or symptom	Immediate action
Acute vertigo + normal head impulse test	MRI-DWI emergency regardless of age
Progressive headache + vomiting + unsteadiness in child	MRI posterior fossa same day
Sudden hearing loss + vertigo	Audiogram + MRI; AICA stroke until proven otherwise
Fever + vertigo + meningism	Blood cultures + empirical antibiotics + CT/MRI before LP
Adolescent + optic neuritis + vertigo	Demyelination likely; MRI brain + spine + neurology
Downbeat nystagmus at primary gaze	Chiari or cerebellar degeneration; MRI posterior fossa

IX. Management Principles and Multidisciplinary Care

X. Summary and Key Clinical Takeaways

Indication	Urgency	Refer to
Any central HINTS sign in acute vertigo	Emergency	Neurology; MRI-DWI within 1 hour
Progressive headache + neurological signs	Emergency	Neurosurgery + oncology if tumour suspected
First demyelinating event	Soon (within 1 week)	Paediatric neurology; MRI brain + spine
Confirmed posterior fossa pathology	Urgent	MDT: neurosurgery/neurology/oncolo

		gy/VRT
Post-treatment vestibular deficit	Routine	Vestibular physician + physiotherapy

References

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I. Introduction: Central vs Peripheral Vestibular Pathology in Children

Central causes of vestibular dysfunction in children are less common than peripheral causes but are disproportionately important because of the catastrophic consequences of missed diagnosis. Posterior fossa tumours are the most common intracranial tumours in children, accounting for approximately 60% of paediatric central nervous system (CNS) neoplasms. Posterior circulation stroke, while rare in children, carries a high risk of mortality and long-term disability if not identified and treated promptly. Demyelinating disease — including acute disseminated encephalomyelitis (ADEM) and multiple sclerosis (MS) — increasingly presents in children and adolescents, with vestibular symptoms as part of a complex neurological syndrome. [1,2]

The challenge of central vestibular pathology in children lies in early recognition. The constellation of vertigo, nausea, gait instability, and nystagmus — which defines the acute vestibular syndrome — can arise from both peripheral and central causes, and the clinical distinction can be subtle. This is particularly true in the emergency setting, where a child with a posterior fossa tumour or stroke may initially appear to have a benign peripheral vestibular illness. Systematic application of central alarm features, combined with a low threshold for neuroimaging, is the cornerstone of safe management. [3,4]

This literature review covers the major central causes of vestibular dysfunction in children — posterior fossa tumours, posterior circulation stroke, and demyelinating disease — as well as rarer central causes including Chiari malformation, syringobulbia, and episodic ataxia. It provides clinicians with a systematic framework for identifying central pathology, selecting appropriate investigations, and coordinating multidisciplinary management.

□ **Important:** A child with acute vertigo AND headache, vomiting, and gait ataxia has a posterior fossa lesion until proven otherwise. Always obtain neuroimaging before attributing symptoms to a peripheral vestibular cause in this clinical context.

Central vs Peripheral Vestibular Feature Differentiation

Feature	Peripheral	Central
Nystagmus direction	Direction-fixed horizontal-torsional	Direction-changing or purely vertical
Head Impulse (HIT)	Positive (corrective saccade)	Normal (no saccade) = ALARM
Skew deviation	Absent	Present = ALARM
Nystagmus change with fixation	Suppressed by fixation	Fixation suppression failure
Course	Acute onset, then gradual resolution	Progressive or failure to improve
Gait ataxia	Mild, directional (falls to lesion side)	Severe, non-directional or truncal
Hearing	Normal or unilateral SNHL (labyrinthitis)	Variable; sudden SNHL + vertigo = AICA alarm
Neurological signs	Absent	May be present

■ Important: In children, trust central alarm features over "normal" HIT.
Normal corrective saccade does NOT exclude central pathology in children.
Any central alarm feature = MRI urgently.

Figure 1. Central vs Peripheral Vestibular Feature Differentiation — key clinical distinctions to guide triage and imaging decisions.
Source: Australian Dizziness Clinics — clinical flowchart.

II. Posterior Fossa Tumours: Classification and Vestibular Presentation

Posterior fossa tumours are the most common intracranial neoplasms in children, with a peak incidence between 5 and 10 years. They account for approximately 60% of all paediatric CNS tumours, compared with only 20% in adults. The posterior fossa contains the cerebellum, brainstem, and fourth ventricle — structures critical for vestibulo-ocular and vestibulo-spinal function. Tumours arising in this space may produce vestibular symptoms directly (by involving the vestibular nuclei, cerebellum, or brainstem pathways) or indirectly (by causing raised intracranial pressure from fourth ventricle obstruction). [1,5]

Medulloblastoma

Medulloblastoma is the most common malignant posterior fossa tumour in children, accounting for approximately 20% of all paediatric CNS tumours. It arises from the cerebellar vermis and typically presents with progressive gait ataxia, truncal instability, and morning headache — the latter reflecting raised intracranial pressure from fourth ventricular obstruction and hydrocephalus. Nystagmus in medulloblastoma is variable in character but may include direction-changing gaze-evoked nystagmus, downbeat nystagmus (from floccular involvement), or positional nystagmus. Papilloedema may be present on fundoscopy — a finding that should never be missed in any child with vestibular symptoms. [1,5]

Ependymoma

Ependymoma arises from the ependymal lining of the fourth ventricle and characteristically extends through the foramina of Magendie and Luschka into the posterior fossa cisterns. Progressive ataxia, vomiting, and cranial nerve palsies (particularly VI and VII) are typical features. The close anatomical relationship between ependymoma and the brainstem vestibular nuclei may produce complex nystagmus patterns and gaze palsy. [1]

Pilocytic Astrocytoma

Cerebellar pilocytic astrocytoma is the most common cerebellar astrocytoma in children and is typically a cystic tumour with a mural nodule arising in the cerebellar hemisphere. It is a WHO grade I tumour with an excellent prognosis following surgical resection. Cerebellar symptoms — including limb ataxia, intention tremor, and gait disturbance — predominate. Nystagmus may be present, particularly gaze-evoked nystagmus toward the side of the tumour. [1]

Brainstem Glioma

Diffuse intrinsic pontine glioma (DIPG) is the most devastating brainstem tumour of childhood — arising in the pons, carrying a near-universally fatal prognosis, and producing a complex syndrome of cranial nerve palsies, pyramidal signs, and cerebellar features. Vestibular symptoms may arise from involvement of the vestibular nuclei in the dorsal pons, producing a diverse range of nystagmus patterns including horizontal gaze-evoked, upbeat, and internuclear ophthalmoplegia (INO). [1,6]

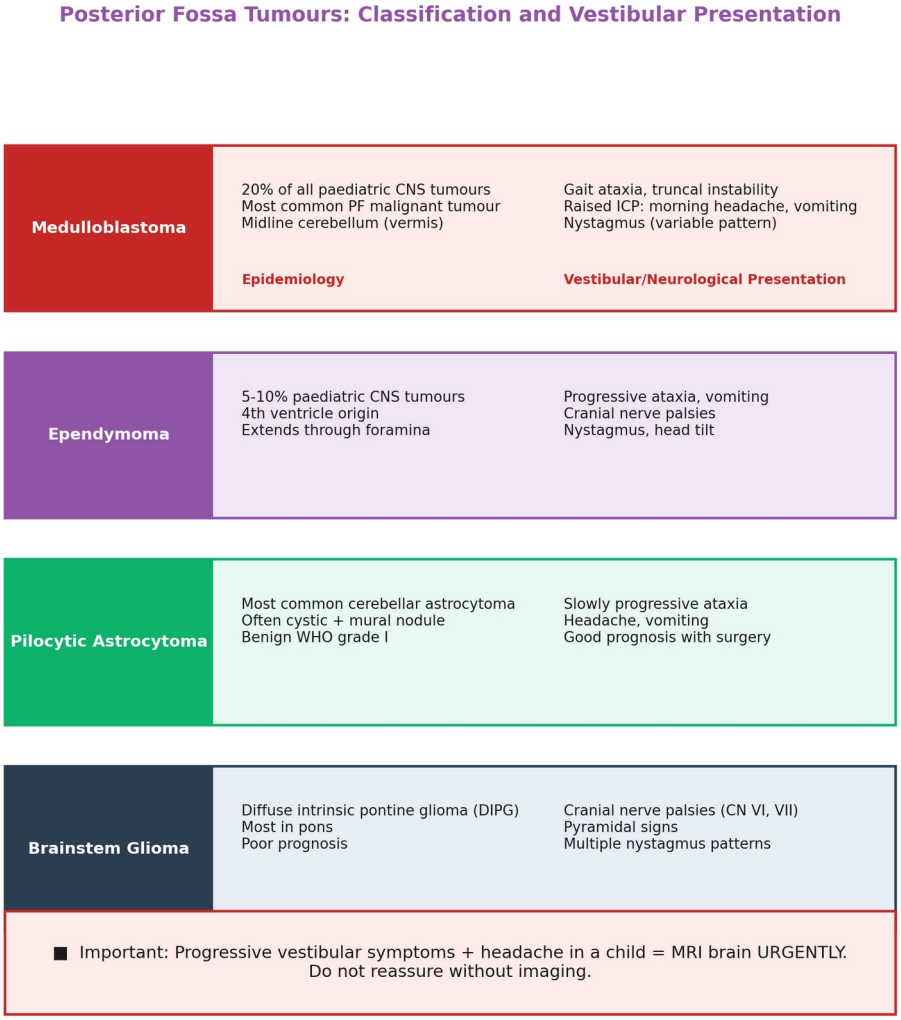

Figure 2. Posterior Fossa Tumour Classification and Vestibular Presentation — epidemiology and key clinical features of the major paediatric posterior fossa tumours.
Source: Australian Dizziness Clinics — clinical flowchart.

□ **Important:** Progressive vestibular symptoms with headache in a child should trigger urgent MRI brain with posterior fossa protocol — never reassure without imaging. Morning headache that worsens with Valsalva or recumbency is a classic feature of raised intracranial pressure from fourth ventricular obstruction.

III. Paediatric Posterior Circulation Stroke

Stroke in children is uncommon but not rare — the overall paediatric stroke incidence is approximately 2–8 per 100,000 per year, with posterior circulation stroke accounting for a significant minority. Posterior circulation stroke in children is particularly challenging because it may present with acute vestibular symptoms indistinguishable from peripheral vestibular neuritis, and because DWI-MRI has a false negative rate of 20–50% in the posterior fossa in the first 24–48 hours. [7,8]

AICA Territory Infarction

Anterior inferior cerebellar artery (AICA) territory infarction involves the lateral pons and — via the labyrinthine artery — the inner ear. The labyrinthine artery is the terminal branch of the AICA and has no collateral supply. AICA infarction therefore produces a distinctive syndrome: acute vertigo, ipsilateral SNHL (cochlear infarction), ipsilateral facial numbness, ipsilateral limb ataxia, and often ipsilateral facial weakness. The cochlear involvement is a critical distinguishing feature from peripheral vestibular neuritis. On HINTS examination, the head impulse test may be abnormal (if labyrinthine involvement produces

canal hypofunction), but central alarm features — such as direction-changing nystagmus or skew deviation — are typically present. [7,9]

PICA Territory Infarction — Wallenberg Syndrome

Posterior inferior cerebellar artery (PICA) territory infarction produces the lateral medullary syndrome (Wallenberg syndrome): vertigo and nausea, ipsilateral facial numbness, contralateral body numbness (spinothalamic crossing), ipsilateral Horner syndrome, dysphonia, dysphagia, and ipsilateral limb ataxia. Nystagmus is typically direction-changing, with a horizontal-torsional pattern that may be complex and atypical. Wallenberg syndrome in children is rare but may occur in the context of vertebral artery dissection following minor head or neck trauma, or with inherited coagulopathy. [7,9]

Basilar Artery Thrombosis

Basilar artery thrombosis is the most catastrophic posterior circulation stroke, producing bilateral brainstem and cerebellar ischaemia. Presentation includes progressive or acute onset of bilateral cranial nerve palsies, impaired consciousness, quadriplegia, and eventual progression to locked-in syndrome if untreated. Early presentation may include vertigo, diplopia, and ataxia. Basilar artery thrombosis in children carries high mortality and demands urgent MRA and neurosurgical/interventional neuroradiology input. [8]

Risk Factors in Children

Unlike adults, paediatric posterior circulation stroke rarely occurs in the context of atherosclerosis or atrial fibrillation. Paediatric risk factors include: congenital cardiac disease with paradoxical embolism; haematological conditions (sickle cell disease, protein C/S deficiency, antiphospholipid syndrome, MTHFR mutation); vertebral artery dissection (post-trauma — including sports, chiropractic manipulation, or minor neck injury); and prothrombotic states. A haematological workup is mandatory in any child with confirmed posterior circulation stroke. [7,8]

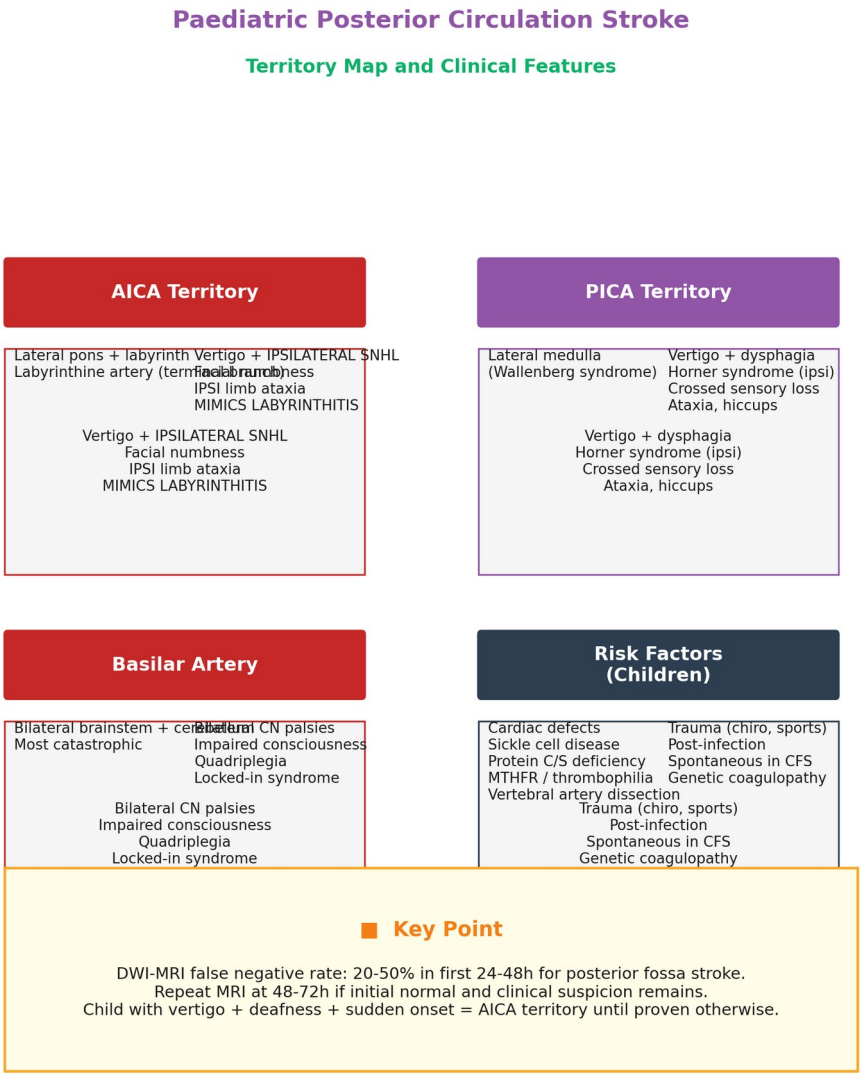

Figure 3. Paediatric Posterior Circulation Stroke — territory map, clinical syndromes, and risk factors for AICA, PICA, and basilar artery territory involvement.
Source: Australian Dizziness Clinics — clinical flowchart.

□ **Clinical Insight:** A child with vertigo + deafness + sudden onset has AICA territory involvement until proven otherwise. The combination of acute vertigo and ipsilateral SNHL mandates urgent MRI regardless of what HINTS examination shows — AICA infarction can produce a positive corrective saccade on HIT if the labyrinthine artery is involved.

IV. Demyelinating Disease: Multiple Sclerosis and ADEM

Demyelinating disease is an important and increasingly recognised cause of central vestibular dysfunction in children and adolescents. Multiple sclerosis (MS), once considered a disease of adults, now has a well-documented paediatric onset — approximately 2–5% of MS patients have onset before age 16, with the majority presenting in adolescence. Acute disseminated encephalomyelitis (ADEM) is a monophasic, predominantly post-infectious demyelinating condition that occurs most commonly in children under 10 years and carries significant acute morbidity. [10,11]

Multiple Sclerosis in Adolescents

Paediatric MS produces relapsing demyelinating plaques that may affect the vestibular nuclei (dorsal pons/medulla), the medial longitudinal fasciculus (MLF — producing internuclear ophthalmoplegia with INO nystagmus), the cerebellar pathways, and the vestibulo-spinal tract. Vestibular symptoms may

include acute onset vertigo, oscillopsia from INO nystagmus, upbeat or downbeat nystagmus from cerebellar/brainstem involvement, or skew deviation. The first demyelinating episode may present as a clinically isolated syndrome (CIS) with vestibular features. [10,12]

Diagnosis of paediatric MS requires application of the McDonald 2017 criteria (adapted for paediatric use), which require dissemination in space and time on MRI plus clinical criteria. CSF oligoclonal bands (OCBs) support the diagnosis. A first episode presenting with vertigo in an adolescent warrants MRI brain and spine, ophthalmological review (for INO and optic neuritis), and paediatric neurology referral before accepting a diagnosis of vestibular neuritis. [10,11]

Acute Disseminated Encephalomyelitis (ADEM)

ADEM is a monophasic, post-infectious demyelinating encephalitis occurring predominantly in children under 10. It is characterised by a polyfocal neurological presentation following viral illness or vaccination, with altered consciousness, pyramidal signs, cerebellar ataxia, and cranial nerve involvement. Vestibular symptoms are common as part of the broader cerebellar-brainstem syndrome. MRI shows multifocal T2/FLAIR hyperintense lesions in white matter, basal ganglia, and brainstem. Management involves high-dose intravenous methylprednisolone. IVIG and plasma exchange are reserved for steroid-refractory cases. [11]

Neuromyelitis Optica Spectrum Disorder (NMOSD)

NMOSD (aquaporin-4 antibody or MOG antibody mediated) can present in childhood with area postrema syndrome (intractable hiccups, nausea, vomiting — caused by dorsal medullary involvement), optic neuritis, and transverse myelitis. The area postrema syndrome may be misdiagnosed as a vestibular or gastrointestinal disorder. AQP4 and MOG antibody testing should be performed in any child with atypical central vestibular symptoms accompanied by nausea, hiccups, or vision loss. [12]

□ **Key Point:** A first demyelinating event presenting with vertigo in an adolescent requires full MS workup — MRI brain and spine, ophthalmological review, and CSF OCBs. Do not assume vestibular neuritis in an adolescent with acute vertigo and any central alarm feature.

V. Other Central Causes: Chiari Malformation, Syringobulbia, Episodic Ataxia

Chiari I Malformation

Chiari I malformation is defined by tonsillar herniation of 5 mm or more below the foramen magnum, producing compression of the lower brainstem and upper cervical cord. In children, Chiari I may present with cough headache (Valsalva-induced occipital headache), downbeat nystagmus (the most characteristic oculomotor sign), central apnoea, and gait instability. Downbeat nystagmus in a child should prompt Chiari MRI even in the absence of symptoms — it is never a normal finding. Symptomatic Chiari I is treated with posterior fossa decompression (suboccipital craniectomy ± C1 laminectomy). [13,14]

Syringobulbia

Syringobulbia is a syrinx (CSF-filled cavity) within the brainstem, typically arising in the setting of Chiari malformation, posterior fossa tumour, or prior trauma/surgery. Involvement of the brainstem tegmentum produces a constellation of findings including horizontal or vertical nystagmus, Horner syndrome (descending sympathetic fibres), palatal weakness, and dysphagia. Diagnosis is by MRI. Management targets the underlying cause. [13]

Episodic Ataxia Type 2 (EA2)

Episodic ataxia type 2 (EA2) is caused by mutations in CACNA1A, encoding the P/Q-type calcium channel alpha-1 subunit. It is the most common episodic ataxia, characterised by attacks of cerebellar ataxia lasting minutes to hours, with prominent vertigo, nausea, and interictal downbeat nystagmus or gaze-evoked nystagmus. Attacks may be triggered by stress, caffeine, exercise, or illness. EA2 typically presents in childhood or adolescence. Acetazolamide (carbonic anhydrase inhibitor) is highly effective in reducing attack frequency and severity in EA2 and should be considered in any child with recurrent episodic vertigo and ataxia. [15]

Episodic Ataxia Type 1 (EA1) and Familial Hemiplegic Migraine

EA1 (KCNA1 mutation) produces briefer attacks (seconds to minutes) of ataxia and vertigo with interictal myokymia — rippling, worm-like muscle movements visible beneath the skin. Familial hemiplegic migraine type 1 (FHM1) is caused by the same CACNA1A mutations as EA2, reflecting allelic variation, and may produce vestibular features as part of the hemiplegic aura. Genetic testing for CACNA1A and KCNA1 should be considered in children with recurrent episodic vestibular symptoms and a family history. [15,16]

□ **Important:** Downbeat nystagmus in a child should prompt MRI for Chiari malformation even if asymptomatic — it is never a normal finding. EA2 (CACNA1A) is the most treatable cause of recurrent episodic vertigo in children: acetazolamide is often dramatically effective.

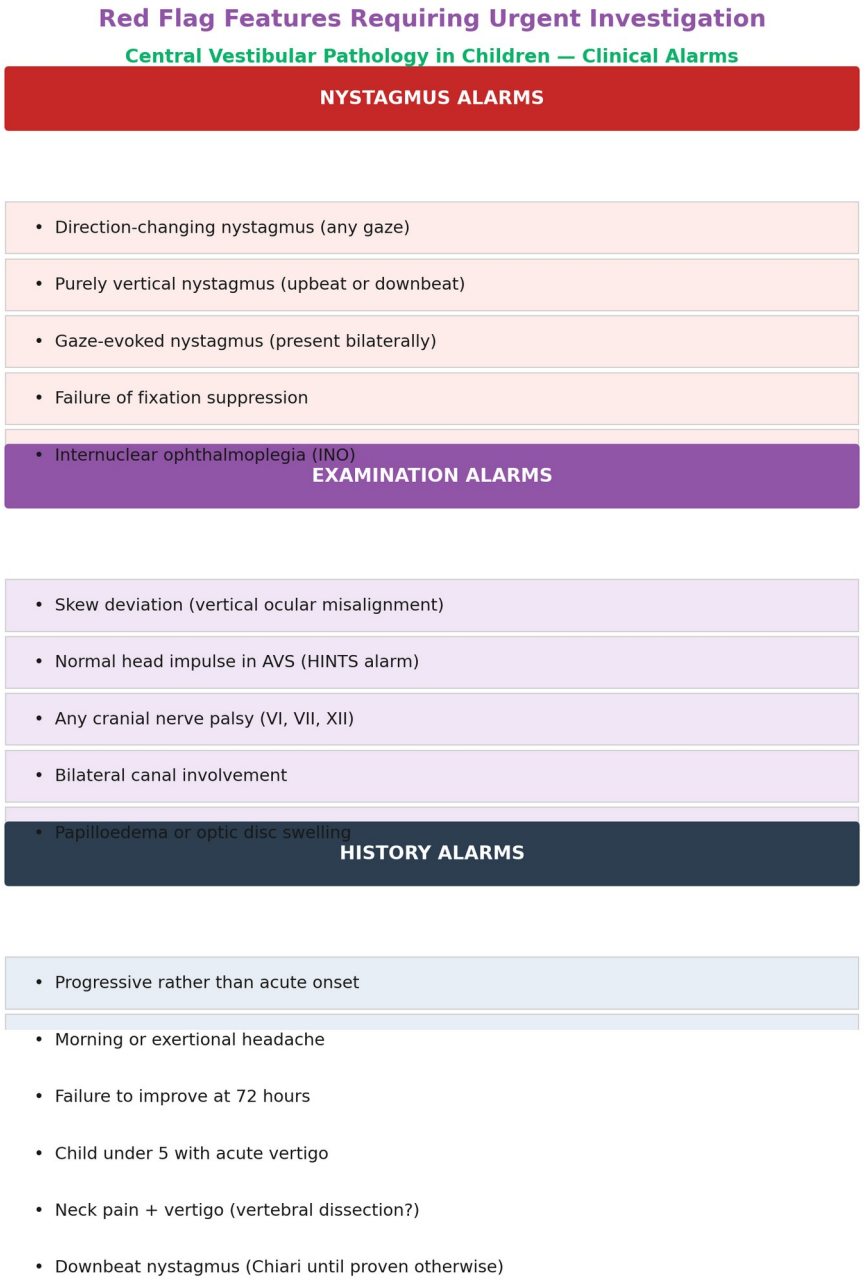

Figure 4. Red Flag Features Requiring Urgent Investigation — central alarm signs categorised by nystagmus, examination, and history features.
Source: Australian Dizziness Clinics — clinical flowchart.

VI. Clinical Features That Point to Central Pathology

Identifying the child with central vestibular pathology requires a systematic clinical approach. The central alarm features can be categorised into oculomotor findings, examination findings, and historical features. The presence of any single central alarm feature warrants urgent neuroimaging. [3,4]

Nystagmus Alarm Features

Direction-changing nystagmus — nystagmus that beats rightward on right gaze and leftward on left gaze (gaze-evoked nystagmus) — is a consistent indicator of central pathology, most commonly cerebellar dysfunction. Purely vertical nystagmus (upbeat or downbeat) is virtually always central in origin. Upbeat nystagmus indicates brainstem lesions (anterior vermis, pontomesencephalic junction). Downbeat nystagmus indicates Chiari malformation, cerebellar degeneration, or drug toxicity. Internuclear ophthalmoplegia (INO) — characterised by adduction limitation on lateral gaze with dissociated abducting nystagmus in the opposite eye — indicates a lesion in the MLF, most commonly demyelination. [3,4]

Examination Alarm Features

Skew deviation (vertical ocular misalignment on the alternating cover test) indicates an otolithic pathway lesion, most commonly in the brainstem or posterior fossa. A normal head impulse test in a child with AVS is a HINTS central alarm sign — it indicates that the labyrinth is functioning normally despite severe vertigo, consistent with a central lesion. The presence of cranial nerve palsies (particularly abducens palsy, facial weakness), pyramidal signs, or cerebellar signs (dysdiadochokinesia, intention tremor) substantially raises the probability of central pathology. Papilloedema is a critical examination finding indicating raised intracranial pressure. [3,17]

Historical Alarm Features

A progressive course — where vestibular symptoms worsen over weeks rather than resolving — is a central alarm. Morning headache, particularly headache that worsens with Valsalva or position change, suggests raised intracranial pressure from posterior fossa mass. Failure to improve by 72 hours in a child initially diagnosed with vestibular neuritis should prompt repeat assessment and consideration of central pathology. Any child under 5 years with AVS requires a low threshold for MRI, given the higher relative incidence of posterior fossa pathology at this age. [4,17]

Central vs Peripheral Vestibular Feature Differentiation

Feature	Peripheral	Central
Nystagmus direction	Direction-fixed horizontal-torsional	Direction-changing or purely vertical
Head Impulse (HIT)	Positive (corrective saccade)	Normal (no saccade) = ALARM
Skew deviation	Absent	Present = ALARM
Nystagmus change with fixation	Suppressed by fixation	Fixation suppression failure
Course	Acute onset, then gradual resolution	Progressive or failure to improve
Gait ataxia	Mild, directional (falls to lesion side)	Severe, non-directional or truncal
Hearing	Normal or unilateral SNHL (labyrinthitis)	Variable; sudden SNHL + vertigo = AICA alarm
Neurological signs	Absent	May be present

■ Important: In children, trust central alarm features over "normal" HIT. Normal corrective saccade does NOT exclude central pathology in children. Any central alarm feature = MRI urgently.

Figure 5. Central vs Peripheral Feature Differentiation Table — systematic comparison of clinical features to guide triage and management.

Source: Australian Dizziness Clinics — clinical flowchart.

□ **Important:** Trust the central alarm features over the HINTS exam in children. A positive corrective saccade on HIT does NOT exclude central pathology in children. Any central alarm feature — nystagmus, examination, or history — mandates MRI.

VII. Neuroimaging in Paediatric Central Vestibular Disease

MRI of the brain and posterior fossa is the definitive investigation for central vestibular pathology in children. The choice of imaging sequences, timing, and whether to include additional studies depends on the clinical context. [7,17]

MRI Sequences and Their Roles

DWI (diffusion-weighted imaging) is the most sensitive sequence for acute ischaemic stroke — demonstrating restricted diffusion within minutes of ischaemia. However, DWI has a false negative rate of 20–50% in the posterior fossa within the first 24–48 hours, due to susceptibility artefacts and the small volume of posterior fossa infarcts. If clinical suspicion for stroke remains after a normal initial DWI, repeat MRI at 48–72 hours is indicated. FLAIR (fluid-attenuated inversion recovery) is the most sensitive sequence for periventricular and juxtacortical demyelinating lesions — the hallmark of MS. Gadolinium-enhanced T1 sequences identify enhancing tumour, abscess, or active demyelinating plaques. CISS or FIESTA sequences provide high-resolution 3D imaging of the labyrinthine structures and internal auditory canals. MRA (magnetic resonance angiography) is indicated when vertebral artery dissection or vascular anomaly is suspected. [7,11,17]

CT Scanning in Posterior Fossa Pathology

CT scanning has limited utility in the assessment of posterior fossa pathology in children. It is significantly inferior to MRI for detecting brainstem and cerebellar pathology due to beam hardening artefact in the posterior fossa. Its primary role in the acute vestibular context is the rapid exclusion of hydrocephalus and obstructive lesions in a child presenting with raised ICP where MRI is not immediately available. CT should not be relied upon to exclude posterior fossa tumour, stroke, or demyelination.

Lumbar Puncture

LP is indicated in suspected demyelinating disease (CSF OCBs for MS diagnosis), CNS infection (bacterial meningitis, viral encephalitis), raised intracranial pressure assessment (opening pressure), and when NMOSD (AQP4/MOG antibody) is suspected. LP should never be performed before CT/MRI when a posterior fossa mass is possible, due to the risk of brainstem herniation.

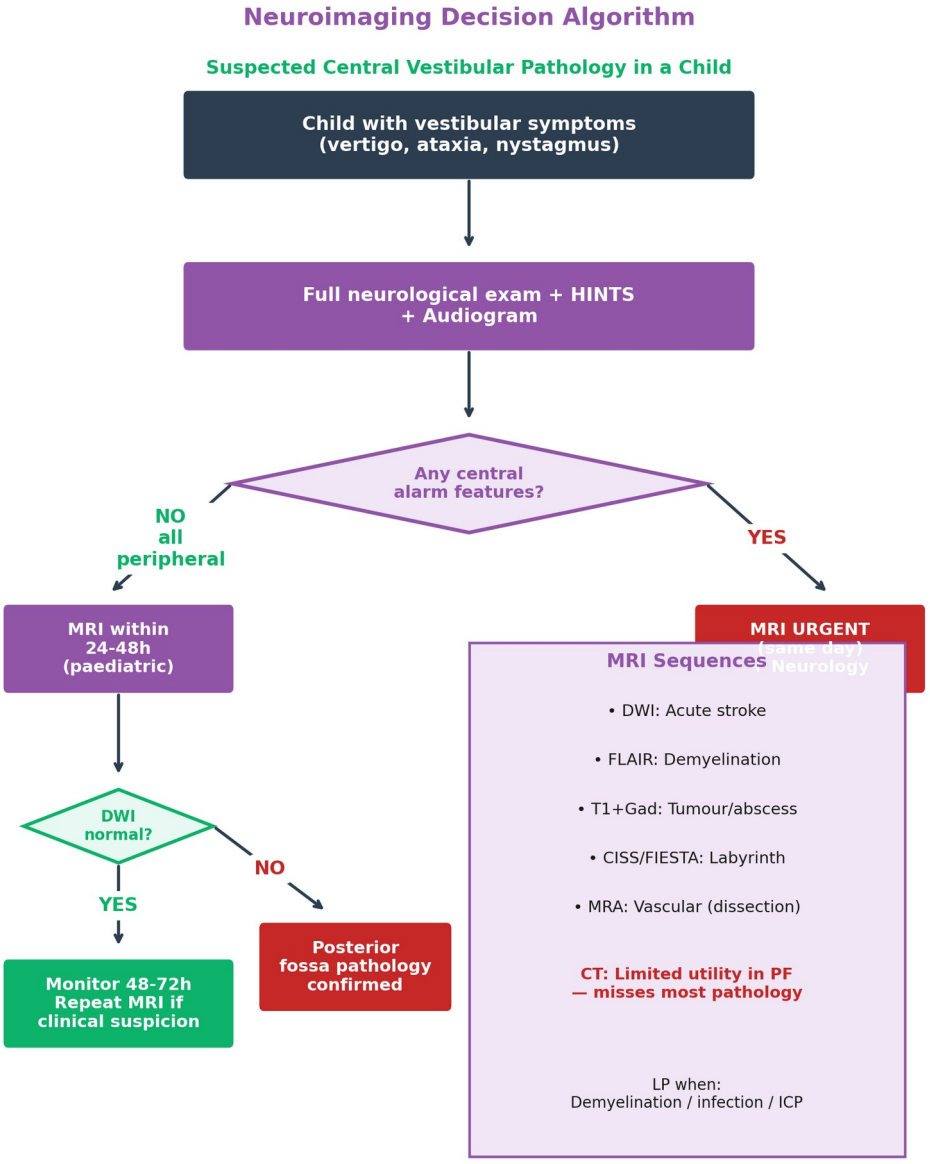

Figure 6. Neuroimaging Decision Algorithm for Suspected Central Vestibular Pathology — sequence selection, urgency stratification, and timing.

Source: Australian Dizziness Clinics — clinical flowchart.

VIII. Vestibular Function Testing in Central Pathology

Vestibular function testing in central pathology differs fundamentally from its role in peripheral vestibular disease. In peripheral disease, vestibular function tests characterise the degree of labyrinthine

dysfunction and guide rehabilitation. In central pathology, vestibular function tests may be normal or show atypical patterns that do not correlate with the clinical syndrome. [18]

Video Head Impulse Test (vHIT)

vHIT tests horizontal SCC function (VOR gain) and is often normal in central pathology — central lesions typically spare individual SCC function, as they affect higher vestibular processing rather than the peripheral sensory organ. A normal vHIT (preserved VOR gain) in a patient with AVS is a central alarm sign on HINTS. However, AICA infarction involving the labyrinthine artery can produce abnormal vHIT — reinforcing that a positive corrective saccade in AVS does not confirm a peripheral cause. [9,18]

Videonystagmography (VNG)

VNG with gaze testing is the most informative vestibular function test for detecting central pathology. Gaze-evoked nystagmus (present bilaterally), rebound nystagmus (on returning gaze to centre), failure of fixation suppression of caloric-induced nystagmus, and optokinetic (OPK) asymmetry are all central signs detectable on VNG. In children, VNG requires adaptation for cooperation and developmental level.

Vestibular Evoked Myogenic Potentials (VEMPs)

Cervical VEMPs (cVEMPs) test the saccular-inferior vestibular nerve pathway through the sternocleidomastoid muscle. Ocular VEMPs (oVEMPs) test the utricular-superior vestibular nerve pathway. Abnormal VEMPs in central pathology reflect involvement of the vestibulo-spinal tract (cVEMP) or the crossed otolith pathway (oVEMP) — patterns that can assist in lesion localisation in brainstem or medullary pathology. [18]

□ **Clinical Insight:** A normal vHIT does not exclude central pathology. In central vestibular disease, always combine vHIT with VNG gaze testing. Gaze-evoked nystagmus on VNG is the most reliable vestibular function test indicator of central pathology. Imaging remains the definitive investigation.

IX. Management Principles and Multidisciplinary Care

Management of central vestibular dysfunction in children is condition-specific and fundamentally multidisciplinary. The immediate priority is accurate diagnosis and appropriate urgency of intervention. [1,2]

Posterior Fossa Tumour Management

Surgical resection is the cornerstone of management for most posterior fossa tumours. Medulloblastoma requires craniospinal radiotherapy and chemotherapy following surgical resection; pilocytic astrocytoma may be cured with complete surgical resection alone. Paediatric neuro-oncology and neurosurgery are the lead specialties. Vestibular rehabilitation following treatment is essential — cerebellar damage from surgery or radiotherapy may produce persistent vestibular and gait dysfunction requiring structured rehabilitation.

Posterior Circulation Stroke Management

Acute paediatric stroke management follows adult principles where evidence-based guidance applies: thrombolysis (tPA) within 4.5 hours if criteria met (growing use in paediatric stroke); endovascular thrombectomy for large vessel occlusion; anticoagulation for vertebral artery dissection. Secondary prevention requires haematological workup and treatment of the underlying prothrombotic condition — including anticoagulation for cardioembolic sources and hydroxyurea for sickle cell disease. Paediatric stroke services and haematology are core MDT members. [7,8]

Demyelinating Disease Management

Acute relapses of MS and ADEM are treated with high-dose intravenous methylprednisolone (typically 30 mg/kg/day for 3–5 days). Plasma exchange and IVIG are reserved for steroid-refractory cases. Long-term disease-modifying therapies (DMTs) for paediatric MS include interferon-beta, glatiramer acetate, and natalizumab — the choice is guided by relapse frequency, MRI burden, and tolerability. Paediatric neurology (MS team) is the lead specialty. [10,11]

Vestibular Rehabilitation in Central Pathology

Vestibular rehabilitation in central vestibular disease uses a different protocol from peripheral disease. In central pathology, substitution strategies (using visual and somatosensory systems to compensate for impaired vestibular function) take priority over habituation (used in BPPV). For patients with complete

unilateral or bilateral vestibular loss from central pathology, visual substitution and body somatosensory retraining are the primary rehabilitation modalities. Gaze stabilisation exercises are appropriate when some VOR function is preserved.

Episodic Ataxia 2 — Acetazolamide

EA2 is one of the most treatment-responsive central vestibular disorders in children. Acetazolamide (5–10 mg/kg/day) typically reduces attack frequency by 50–75% and may completely abolish attacks in some patients. The mechanism likely involves modulation of potassium and bicarbonate flux in cerebellar neurons. Monitoring for nephrolithiasis and metabolic acidosis is required with long-term acetazolamide therapy. 4-aminopyridine is an emerging alternative for EA2. [15]

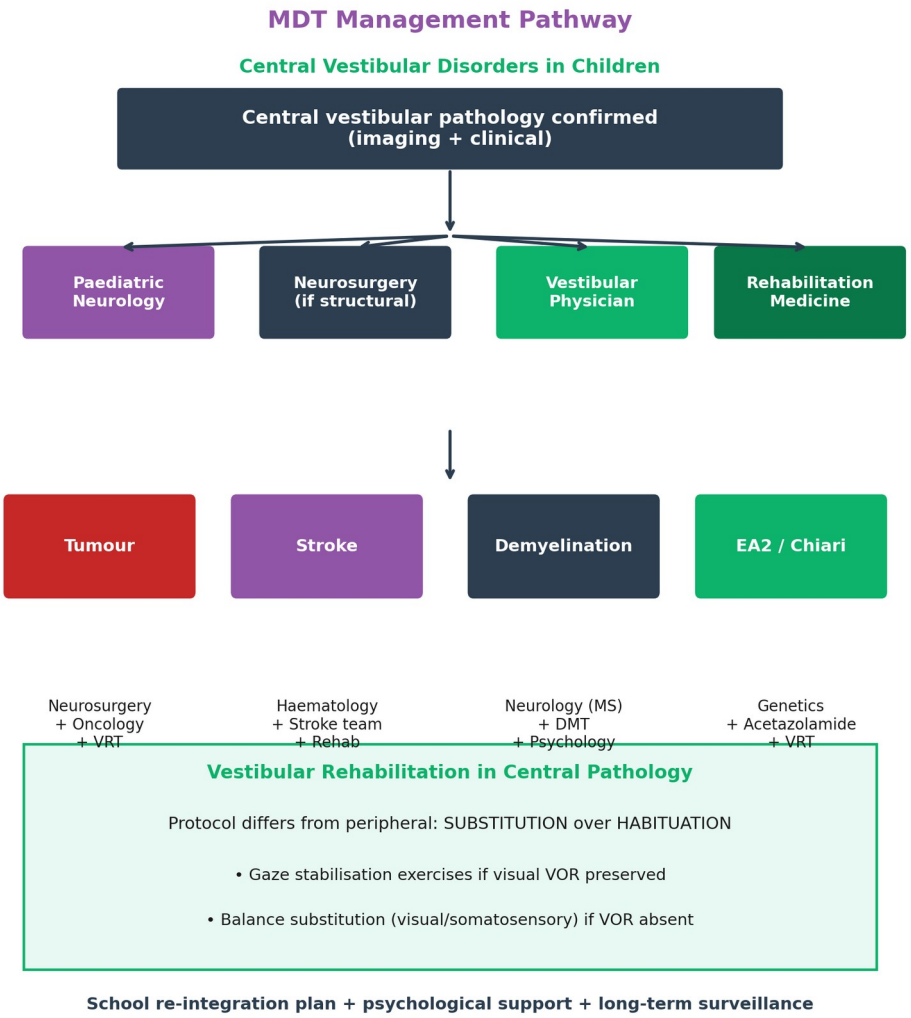

Figure 7. MDT Management Pathway for Central Vestibular Disorders in Children — condition-specific teams, vestibular rehabilitation, and school re-integration.
Source: Australian Dizziness Clinics — clinical flowchart.

□ **Key Point:** School re-integration planning and psychological support are essential components of management for children with central vestibular disorders. Prolonged absence from school compounds the psychosocial burden of chronic neurological illness. Early involvement of school liaison nurses and educational psychologists improves long-term outcomes.

X. Summary and Key Clinical Takeaways

Central causes of vestibular dysfunction in children span a broad spectrum from benign treatable conditions (EA2, Chiari I) to immediately life-threatening emergencies (basilar artery thrombosis, medulloblastoma with hydrocephalus). The following ten points encapsulate the core clinical principles:

1. Posterior fossa tumours are the most common intracranial tumours in children — always consider them in any child with progressive vestibular symptoms or headache.

2. Morning headache, vomiting, and papilloedema = raised intracranial pressure from posterior fossa mass until proven otherwise. MRI is mandatory.

3. A child with vertigo + ipsilateral SNHL = AICA territory stroke until proven otherwise. MRI-DWI urgently, even if HINTS appears peripheral.

4. DWI-MRI has a 20–50% false negative rate in posterior fossa stroke in the first 24–48 hours. Repeat at 48–72 hours if suspicion remains.

5. Vertebral artery dissection is a cause of paediatric posterior circulation stroke — ask about trauma, sports, neck manipulation.

6. ADEM in a child presents with polyfocal neurological features post-infection — multifocal MRI lesions and altered consciousness distinguish it from MS.

7. A first demyelinating episode presenting with vertigo in an adolescent warrants full MS workup before attributing to vestibular neuritis.

8. Downbeat nystagmus is never normal in a child — always obtain MRI for Chiari malformation.

9. EA2 (CACNA1A) is the most treatable cause of recurrent episodic vestibular symptoms in children — acetazolamide is often dramatically effective.

10. Central vestibular rehabilitation uses substitution (not habituation) as the primary strategy — refer early to a vestibular physiotherapist familiar with central protocols.

This review is the sixth in the Vestibular Medicine in Children series. The next review (PVM07) covers Enlarged Vestibular Aqueduct Syndrome — genetics, pathophysiology, audiological management, and vestibular implications.

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