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About the Journal

Aims and Scope

The *Sri Lanka Journal of Neurology (SLJoN)* is the official journal of the Association of Sri Lankan Neurologists (ASN) The journal aims to facilitate the sharing of new evidence and the promotion of research in the field of neurology by providing a platform for publication for all health professionals interested in Clinical Neurology. Neurosurgery and Neurosciences. It is an open access, peer-reviewed journal.

The scope of the journal includes dissemination of new evidence, encouragement of debate on current evidence and practice, exchange of ideas to promote good clinical practice and influence on policy making related to health care in Neurology and Neurosciences.

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The inaugural issue of the *SLJoN* was published in 2012, with Prof. Saman Gunatilake as the Editor, a 7-member Editorial Board and 6-member International Advisory Board. Since then, it has been published annually.

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EDITORIAL

Artificial intelligence and academic writing

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Artificial intelligence revolution is here to stay and all of us use it to varying degrees. One recent survey in UK has found that 92% of university students use AI, and 20% have used AI for their assignments. While such use can improve the learning experience it also can have its drawbacks. Widespread availability of AI tools like ChatGPT, Google Gemini, Claude have led to its use by students in their academic work, teachers in their teaching and scientists in their research. AI enhances academic writing in six areas: idea generation, content structuring, literature synthesis, data management, editing, and ethical compliance. AI has also impacted scientific journals in a big way positively and also negatively. Some of the articles submitted even to our journal sometimes raise our concern whether it's AI generated or plagiarized. To detect these, journal publishers and editors use AI tools and now the SLJoN too has access to "ithenticate" one of the trusted plagiarism checker and AI content detector.

Artificial intelligence (AI) is the capability of computational systems to perform tasks typically associated with human intelligence, such as learning, reasoning, problem-solving, perception, and decision-making. AI can produce text, improve writing style, and also analyze data. It also can perform language editing and check and correct references. AI also can produce entire articles like reviews on request. It can be difficult to recognize original work written by an individual and work generated by AI. This has become a serious problem in the academic world though some uses of AI are seen as acceptable.

There are two categories of AI use in academic writing – AI-assisted content and AI-generated content. AI-assisted content is work that is predominantly written by the authors but has been improved with the aid of AI tools. Using AI to assist with grammar checks, enhance sentence clarity are generally acceptable to the editors and publishers. The author is in control, and the AI is used merely as a tool to polish the final product. There is no need for formal disclosure and the work has to be original. AI-generated content is produced almost totally by the AI itself. The AI tool generates significant portions of text, or even entire sections, based on detailed instructions (prompts) provided by the author. AI generated content may have plagiarized someone's work or infringed on copyright. This type of content may face restrictions. Publishers may even reject it.

Guidelines (Sage and the Committee on Publication Ethics) are now available on the use of AI in academic work. All the guidelines say AI tools can be used for routine tasks like improving grammar, assisting with literature searches and these applications do not require specific acknowledgement. AI generated content is not allowed unless there are clear reasons why this was necessary for the research. Depending on how AI is used, it must be referenced in the manuscript.

AI generated fake papers are another major hazard. These are AI-generated rewrites of previously published studies. The structure, methods and even figures and tables closely mirror those of the originals, but the papers actually have abnormally low text similarity scores. While genuine manuscripts typically show about 10-15 per cent similarity in ithenticate, these fakes register only around 2-5 per cent. Those low scores are in part accounted for by the awkward or vague phrasing they contain, replacing commonly accepted scientific terms with less precise expressions. Some references appear to be inserted at random, and some are unrelated to the study's topic.

The use of AI can improve efficiency and productivity. It has the ability to create well-written and elegant articles for scientific journals. Yet, researchers and authors cannot be complacent in using this tool; instead, they must be fully accountable for their work and must actively engage in the output created.

Saman Gunatilake

Joint Editor

SLJoN

REVIEW

Restricting valproate prescribing in women – path forward in Sri Lanka

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Abstract

Valproate is a well-established antiseizure medication (ASM) with efficacy across multiple seizure types. However, its teratogenic effects and risks to neurodevelopment in exposed children present significant challenges in prescribing it to women of childbearing potential (WOCP). Various countries have implemented stringent guidelines to limit valproate's use among women. This article examines the pharmacological profile, efficacy, and reproductive health risks of sodium valproate, reviews international restriction practices, and proposes actionable steps for Sri Lanka.

KEYWORDS

Valproate (VPA), Anti-seizure medication (ASM), Women of childbearing potential (WOCP), Major congenital malformation (MCM)

INTRODUCTION

Globally, an estimated 5 million people are diagnosed with epilepsy each year. In high-income countries, there are estimated to be 49 per 100 000 people diagnosed with epilepsy each year. In low- and middle-income countries, this figure can be as high as 139 per 100 000.[1]

Sri Lanka is a middle-income country in Southeast Asia with a population of approximately 22 million. Its state care health system provides free health to all its citizens. Epilepsy is treated both in the private and government health sector. Anti-seizure medications (ASMs) are prescribed by junior doctors to consultants without restriction.

Women with epilepsy are at a higher risk of teratogenic effects of ASM. ASMs associated with highest teratogenicity are sodium valproate and topiramate. Drugs with the least teratogenic effects include lamotrigine, levetiracetam, oxcarbamazepine and have a lower risk than that associated with the lowest dose category of valproate.[2]

Valproic acid was first approved for the treatment of epilepsy in France in 1967. However, the first controlled trial documenting its efficacy was published in 1975.[3] The recent discovery of its ability to inhibit histone deacetylase has rekindled interest in novel potential indications, such as cancer. It is the conventional drug of choice for adults and children with a clear diagnosis of genetic generalized epilepsy, otherwise known as idiopathic generalized epilepsy (IGE).[4] Since its approval for the treatment of epilepsy decades ago, the clinical indications for valproic acid have evolved to include migraine prophylaxis and treatment of bipolar disorders.

In one study in the USA, patients with migraine and mood disorders accounted for the largest proportion of valproic acid use and had the highest pregnancy rates, while patients with epilepsy had the lowest. This is despite restrictions in the USA.[5]

Valproate, discovered in the 1960s, has been a cornerstone in epilepsy management due to its broad-spectrum efficacy,

especially in generalized epilepsies. However, evidence of serious teratogenicity, including structural birth defects and neurodevelopmental disorders, warrants caution when prescribing sodium valproate to women of childbearing potential.

In response, many countries have implemented regulatory frameworks to limit its use among young females. This article assesses the necessity for similar measures in Sri Lanka.

Sodium valproate: Overview and mechanism of action

Sodium valproate has multiple mechanisms of action, including GABA potentiation, blocking of T-type calcium channels and blocking of sodium channels. This effect stabilizes neuronal firing and is beneficial across a variety of seizure types. Valproate is enzyme inhibitor and extensively metabolized by conjugation and oxidation. The half-life in adults is between 13 to 16 hours.[6]

Valproate has a broad spectrum of efficacy against all focal and generalized seizures. However, it is more effective in generalized tonic-clonic seizures, absence seizures, myoclonic seizures, and some mixed seizure types. It is less effective in focal seizures, where alternative ASMs may offer more targeted control.

Valproate is associated with a dose-related teratogenicity rate higher than any other marketed antiseizure medication, with risk of major malformations higher than 30% at doses greater than 1100 mg/d. In utero exposure is also associated with dose-dependent reduced verbal IQ, other cognitive dysfunction, and autism.

Adverse effects on reproductive health in females

The reproductive risks associated with sodium valproate are well-documented with major congenital malformations (MCM) and neurodevelopmental risks.

Major congenital malformation (MCM)

Around 1 in 9 babies (11%) will have a birth defect in women who take valproate while pregnant. In the first trimester valproate increases the risk of major congenital malformations, independent of any contribution of the epilepsy syndrome itself.

Birth defects seen when mothers take valproate during pregnancy include neural tube-like defects (e.g., spina bifida, open lumbosacral myelocle) in 1 to 2 percent of fetuses, which represents a 10- to 20-fold increase over the general population.

Additional patterns of major malformations associated with first-trimester valproate exposure include oral clefts, cardiovascular and urogenital malformations, and multiple malformations (Table 1).[7]

The risk of MCMs during valproate treatment is highest during the first 7 weeks of pregnancy and the rates of MCMs are dose dependant (Table 2).[8]

The reasons for the teratogenic effects are not fully understood, but possibly involve epigenetic effects, including the inhibition of histone deactylase with associated changes in gene expression, increases in foetal oxidative stress, or the antagonism of folate required for DNA synthesis.[9]

TABLE 1 Odds ratios and absolute risk of congenital malformation with sodium valproate (adapted from Jentink et al., 2010)[7]

	Odds ratio (median and range) in offspring of mothers who took valproate in pregnancy	Absolute risk
Spina bifida	12.7 (7.7-20.7)	0.6%
Atrial septal defect	2.5 (1.4-4.4)	0.5%
Cleft palate	5.2 (2.8-9.9)	0.3%
Hypospadias	4.8 (2.9-8.1)	0.7%
Polydactyly	2.2 (1.0-4.5)	0.2%
Craniosynostosis	6.8 (1.8-18.8)	0.1%

TABLE 2 Prevalence of major congenital malformations (MCMs) in offspring exposed prenatally to valproate monotherapy

ASM treatment (dose range, mg/d)	Prevalence of MCMs (95% CI), %
Valproate (≤650)	6.0 (4.4-8.0)
Valproate (>650-≤1450)	11.1 (8.9-13.6)
Valproate (>1450)	25.2 (17.8-33.8)
Valproate (100-3000)	9.9 (8.5-11.5)

Neurodevelopmental risks

Of all ASMs, valproate is the drug most strongly associated with adverse neurodevelopmental outcomes. In women who take valproate while pregnant, about 3-4 children in every 10 may have developmental problems, and these disorders can be seriously debilitating and permanent.[10] Unlike major congenital malformations, antiseizure medication exposure during the last trimester may be the most detrimental for cognitive performance.[11]

Cognitive impairment

Several publications have described an association between fetal exposure to valproate and the risk of impaired IQ and cognitive developmental delay. One study found a significantly lower IQ (mean IQ score of 97) in children of women with epilepsy treated with valproate than with lamotrigine (mean IQ score of 108) and these findings persisted at six years. The effects are dose dependent with valproate doses above 800-1000 mg/day have been consistently associated with reduced IQ with doses below the 800 mg/day threshold have been linked to worse verbal IQ performance and a need for educational support.[12]

Unlike major congenital malformations, antiseizure medication exposure during the last trimester may be the most detrimental for cognitive performance.[12]

Behavioural impairment

Valproate has been consistently associated with a 2- to 4-fold increased risk of autism spectrum disorder (ASD), 2- to 5-fold increased risk of intellectual disability (ID), and poor adaptive functioning.[13]

Studies have shown that the risk of autism spectrum disorder with valproate exposure was 4.4%, compared to 1.5% in the general population. In another study, the correlation of autism spectrum disorder to fetal antiseizure medication exposure was not seen in children exposed to polytherapy without valproate, suggesting that valproate (or valproate dose) rather than polytherapy is the critical determinant of the relationship.[14,15] In addition, children with in utero exposure to valproate are also at a significantly greater risk for a diagnosis of attention deficit hyperactivity disorder (ADHD).[16]

Rationale for restricting sodium valproate in reproductive-aged females

Restricting valproate use in young, healthy females is crucial to prevent potential teratogenic effects and neuro-developmental issues in exposed children. Given the availability of alternative ASMs with lower teratogenic risks, a more selective approach in prescribing valproate can help protect the health of both women and potential offspring.

Unplanned pregnancy while on valproate: what can be done?

Pregnancy while taking valproate presents significant challenges and requires careful management based on a risk-benefit assessment. Poor seizure control during pregnancy poses substantial risks to both the mother and baby, underscoring the importance of individualized care. The priority in the first trimester should probably be to withdraw the drug or reduce the dose to prevent cognitive impairment in the newborn.[17] If a switch is decided, levetiracetam is preferable to lamotrigine, as it can be quickly loaded, either orally or intravenously.

The evidence is unclear whether to switch to another ASM while pregnant, as switching form valproate to another ASM is associated with risk. One study had shown that generalised tonic-clonic seizures were twice as common in those where valproate was withdrawn (33%; n=93) or replaced (29%; n=38) in pregnancy, compared with those

who stayed on it (16%; n=1588). This needs to be kept in mind while considering the risk vs benefit of switching to another ASM during pregnancy.[18]

In some circumstances, you may not be able to switch to another medication. In such cases, the pregnancy should be closely monitored (e.g., with high-resolution ultrasound). The consequences of any MCM detected should be discussed in depth and women should be offered counselling to help them take the best possible decision.

Prescription of valproate in paediatric age group

Initiation of valproate before menarche is not recommended, although there are circumstances when it might be acceptable. Valproate can be prescribed as first-line therapy for potentially self-limited epilepsy syndromes, such as focal epilepsy with centrotemporal spikes, childhood absence epilepsy, and benign myoclonic epilepsy in infancy.[19]

It should not be the first choice in patients with chronic childhood-onset epilepsy syndromes likely to remain active into adolescence and adulthood (e.g., Jeavons syndrome). If starting valproate, adequate information must be given to both patients and their parents/legal guardians regarding future implications.

Contraception while on valproate

Contraception in women of childbearing potential (WOCP) with epilepsy on valproate is crucial and family planning is important. WOCP on valproate should use at least one effective method of contraception (preferably a user-independent method such as an intrauterine device or implant) or two complementary methods including a barrier method.

Pregnancy planning while on valproate

Pregnancy should be planned approximately one year in advance to allow sufficient time to safely withdraw valproate and find an effective alternative. The minimum period recommended for switching from valproate to any other ASM is 3 months.

As a general recommendation, contraception should be maintained for three months after valproate discontinuation to ensure full clearance and allow time to adapt to the new ASM. Folic acid supplementation is recommended for at least one month before conception, regardless of valproate treatment. The recommended dose for patients taking ASM is 4-5 mg/day, which is higher than doses recommended for women at low risk.[20]

Switching valproate in focal epilepsy and generalized epilepsy

Sodium valproate has not shown to be superior to other ASM in focal onset epilepsy.

For generalized epilepsy syndromes

In those with epilepsy with GTCS, monotherapy with lamotrigine and levetiracetam should be tried first, followed by combination therapy with the two drugs if necessary. For those with juvenile myoclonic epilepsy, levetiracetam, followed by lamotrigine, is the first-line alternative to valproate. Combination therapy with lamotrigine and levetiracetam can be considered. Ethosuximide, followed by lamotrigine, should be the first option for childhood absence epilepsy. If absence seizures persist, levetiracetam should be tried. The first-line options for juvenile absence epilepsy are lamotrigine and levetiracetam. Ethosuximide can be considered as a later alternative.

Switching from valproate to lamotrigine

Switching from valproate to lamotrigine requires on average 2-3 months or longer.

LTG dose starting at 12.5mg with dose escalation every two weeks with the aim to reach target dose in 8 weeks followed by sodium valproate withdrawal over six to eight weeks.

Switching from valproate to levetiracetam

Switching from valproate to levetiracetam is faster, as titration is not always needed

The target dose for levetiracetam is between 1000 and 2000 mg/day depending on the valproate dose. Valproate should not be decreased abruptly due to the risk of breakthrough GTCS. Selection of these alternatives should be based on seizure type, individual response, and safety profile.[4]

Situations when valproate could be continued

Focal onset epilepsy

In self-limited focal childhood epilepsy syndromes such as focal epilepsy with centrotemporal spikes) valproate should be both started and stopped before the patient reaches reproductive age. Four ASMs can be recommended as alternatives for focal epilepsy based on known risks of MCMs from the EURAP registry. These are levetiracetam, lamotrigine, oxcarbazepine, and carbamazepine.

Generalized epilepsy

Valproate may be continued in specific situations where its benefits outweigh the risks. These include cases where it is the only effective medication after other antiseizure medications (ASMs), whether as monotherapy or polytherapy, have failed, or when alternative ASMs are contraindicated due to severe psychiatric disorders or allergies. It may also be considered when seizures are expected to resolve, such as in childhood absence epilepsy, benign myoclonic epilepsy in infancy, or when the patient is a candidate for epilepsy surgery. Additionally, valproate may be used in severely disabled females with little to no likelihood of pregnancy or when the risk of seizure exacerbation is deemed more critical than the potential teratogenic effects.

How can we minimize the risks for women who require valproate after attempting alternative treatments?

To minimize risks in women who require valproate after considering and trying alternatives, several measures can be implemented. Valproate should be prescribed at the minimum effective dose, preferably no higher than 600-800 mg/day, while avoiding high peak concentrations by utilizing prolonged-release formulations and fractionating the dose. High-dose folic acid (4-5 mg/day) is recommended before conception and should be continued for at least the first three months of pregnancy to reduce the risk of neural tube defects. Additionally, if valproate cannot be completely withdrawn, combining it in polytherapy with the lowest possible dose is preferable to using higher-dose monotherapy. These strategies aim to balance the need for seizure control with minimizing potential adverse effects.[21]

International guidelines and restriction models

Many countries have introduced stringent measures to control the prescription of sodium valproate for women of childbearing potential due to its high teratogenic risk and potential for neurodevelopmental harm in children exposed in utero.

In the European Union (EU), a Pregnancy Prevention Program (PPP) mandates that healthcare providers offer educational materials to patients and providers, obtain signed risk acknowledgment forms, and limit valproate prescriptions to specialists, who must conduct annual reviews and provide contraceptive counselling.[22]

Since 2018, in the UK valproate is not to be used in any female able to have children unless there is a Pregnancy Prevention Programme (PPP) in place. This is designed to make sure patients are fully aware of the risks and the need to avoid becoming pregnant. Healthcare professionals who seek to prescribe or dispense valproate to their female patients must make sure this is within the terms of the PPP. This includes the completion of a signed risk acknowledgement form when their treatment is reviewed by a specialist, at least annually.[23] These regulatory changes are further supported by smaller pack sizes to encourage monthly prescribing and a pictogram or warning image on valproate labelling. Regulations introduced in 2023 are to ensure all patients receive the whole pack of valproate with warnings on the box.

The United States Food and Drug Administration (FDA) has issued a boxed warning emphasizing that valproate should not be administered to women of childbearing potential unless other medications have failed to provide adequate symptom control or are deemed unacceptable, however it stops short of mandating annual reviews or signed acknowledgment forms.[24]

Australia's Therapeutic Goods Administration (TGA) has adopted a Pregnancy Prevention Program akin to the EU's, restricting prescribing to specialists and providing risk acknowledgment forms and educational materials to improve patient understanding.

In Southeast Asia (e.g., Malaysia, Singapore)

In Malaysia, the national pharmaceutical regulatory agency (NPRA) has issued warnings about valproate's teratogenicity and have contraindicated valproate use in epilepsy and bipolar affective disorder if no PPP (pregnancy prevention program) is in place. In addition, healthcare providers are urged to ensure informed decision-making by patients, with a focus on contraception use.[25] In Singapore too, valproate is contraindicated during pregnancy unless no alternative is available. Women on valproate are advised to use effective contraception, and regular reviews are conducted. These regulatory frameworks have helped reduce the incidence of foetal valproate exposure. There are no legal restrictions placed in our neighbouring countries in India, Pakistan, Bangladesh, Maldives, Bhutan, Nepal and Myanmar.

Current challenges in Sri Lanka

In Sri Lanka, several unique challenges complicate the implementation of valproate restrictions.

1. Limited awareness: Among both healthcare providers and patients, there is limited awareness of valproate's teratogenic risks. Inadequate counselling facilities further exacerbate the problem, leaving many women unaware of the associated risks.

2. Resource constraints: Sri Lanka faces significant resource constraints in access to alternative anti-seizure medication, particularly newer-generation options.

3. Sociocultural factors: Discussing contraception remains a sensitive topic in Sri Lanka. Women may be reluctant to disclose their reproductive plans, and healthcare providers may hesitate to engage in these discussions due to cultural norms.

4. Reluctance of the healthcare providers to change their lifelong practises with emerging new evidence.

The path forward for Sri Lanka

Given the international precedent and the known risks associated with sodium valproate, Sri Lanka should consider adopting a structured approach to minimize its use among women of childbearing potential. Firstly, and most importantly we need to support and develop a pregnancy registry documenting birth, and developmental defects associated with antiseizure medications during pregnancy.

These are some of the proposals which could be considered for implementation.

1. **Developing national guidelines:** Create guidelines specifically addressing the use of valproate in women of childbearing potential, including recommendations for alternative treatments.

2. **Educational programs for healthcare providers:** Train healthcare providers on the reproductive risks associated with valproate and encourage the discussion of safer alternatives with patients.

3. **Informed consent and mandatory counselling:** Implement a formal consent process where prescribers are required to discuss the risks with patients and document consent.

4. **Restricting prescription to specialists:** Limit initial valproate prescriptions to specialists in neurology and epilepsy, with a focus on informed decision-making.

5. **Alternative ASM Access:** Ensure availability and affordability of alternative ASMs, such as lamotrigine and levetiracetam, which have lower teratogenic risks.

6. **Discourage** use of sodium valproate for migraine and bipolar affective disorder when suitable alternatives are available.

7. **Other options** such as smaller pack size to encourage monthly prescriptions and pictural warning for valproate.

Sri Lanka's health sector also should be mindful and should implement practical solutions to restrict valproate use in women of reproductive age. This is essential to prevent major congenital malformations (MCM) and adverse neurodevelopmental outcomes, which place a significant burden on individuals, families, and healthcare facilities already under severe economic constraints. To adopt the above, we would need consensus among stakeholders and a national policy based on evidence.

CONCLUSION

Restricting sodium valproate prescribing among females of reproductive age in Sri Lanka is an essential step to reduce teratogenic risks and improve reproductive health outcomes. Implementing guidelines for limited and well-informed valproate use, combined with educational initiatives and availability of alternative ASMs, can contribute to a safer, more effective approach in epilepsy management for this demographic.

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ORIGINAL RESEARCH

A clinical audit on febrile seizure management at the primary care unit of Lady Ridgeway Hospital for Children, Colombo

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Abstract

Introduction: Febrile seizures (FS) are the most common childhood seizures, categorized into simple and complex types. Simple febrile seizures (SFS), which are benign and self-limiting, account for over 70% of cases and often prompt emergency visits, sometimes leading to unnecessary investigations. Data on adherence to FS management guidelines in Sri Lanka are scarce.

Objectives: To evaluate adherence to standard FS management guidelines at the Primary Care Unit (PCU), Lady Ridgeway Hospital (LRH), Colombo.

Methods: A prospective clinical audit was conducted at LRH PCU from August 1-30, 2023, including 25 FS cases via convenience sampling. Data were collected by direct observation using a checklist. Parental counselling was assessed using an expert-validated eight-point guide.

Results: The mean age was 2.63 years, and 56% were male. They presented due to SFS in 68% and complex FS in 32%. There were two children with febrile status epilepticus. URTIs were the most common cause of fever (56%). First-time seizures accounted for 66% of admissions. Over-investigation was observed in 65% of SFS; 9.5% of eligible patients were under-admitted. Buccal midazolam (IV preparation, off-label) was the most frequently used first line.

Parental counselling was inconsistent and inadequate. It was as verbal instructions in 32% and in written form in 16%. Key educational points were incompletely addressed; epilepsy risk was never discussed. However, inappropriate prescription of intermittent prophylaxis (clobazam) was minimal.

Conclusion: The audit revealed over-investigation, under-admission, and inadequate parental counselling, highlighting the need for standardized protocols and structured caregiver education.

KEYWORDS

Seizures, Febrile; Guideline Adherence; Primary Health Care; Health Education; Anticonvulsants

INTRODUCTION

Febrile seizures, or commonly referred to as "febrile fits," are a frequent medical concern, especially in the paediatric population. According to the International League Against Epilepsy (1993), a febrile seizure is defined as "an epileptic seizure occurring in childhood after age of one month, associated with a febrile illness not caused by an infection of the central nervous system (CNS), without previous neonatal seizures or a previous unprovoked seizure, and not meeting criteria for other acute symptomatic seizures".[1]

There are two distinct clinical presentations: simple and complex febrile seizures. Simple febrile seizures, accounting for more than 70%, are typically generalized, last less than 15 minutes, and occur once in a febrile illness. Complex febrile seizures, which constitute the remaining 30%, may last longer than 15 minutes, recur within a single febrile illness, or present with focal features.

Febrile seizures affect approximately 2-5% of the at-risk paediatric population,[2] making them one of the most common neurological emergencies in children and a frequent cause of emergency medical evaluation. Ensuring that clinical practice aligns with standard guidelines is critical in maintaining high-quality pediatric care. Despite being usually benign, febrile seizures often induce considerable anxiety among caregivers, leading to emergency visits and extensive evaluations. Local studies suggest persistent parental misconceptions regarding the nature and management of febrile seizures.[3] This highlights the importance of comprehensive discharge education to ease fears and provide practical home management strategies.

The aim of this audit was to evaluate the current clinical management of febrile seizures at the Primary Care Unit (PCU) of the leading paediatric hospital in the country, assess adherence to national and international guidelines, and identify key areas for improvement in order to enhance the quality of care provided.

Audit standards

The PCU primarily adheres to "The National Guidelines for Management of Seizures" and evidence from the "Management of Simple Febrile Seizures; *Sri Lanka Journal of Child Health*, 2017."Acute seizure management is guided by the Advanced Paediatric Life Support (APLS) protocols. These sources provided the standard for comparison with existing practices.

METHODS

A prospective audit was conducted on a convenient sample of children presenting with febrile seizures to the PCU at Lady Ridgeway Hospital for Children over a one-month period, from 1st to 31st August 2023. Ethical clearance was granted by the Ethics Review Committee, Faculty of Medicine, University of Colombo. The audit was observational, with no patient or parental interviews conducted. Management practices were assessed through direct observation by auditors using a structured checklist. Parental advice was evaluated qualitatively with the aid of an expert-validated eight-point assessment guide.

RESULTS

A total of 25 children were assessed during this month, comprising 14 males (56%) and 11 females (44%), with a mean age of 2.63 years (SD: 2.22). One child was an outlier, being outside the typical 6 months to 6 years age range.

Regarding seizure characteristics, 68% (17 children) experienced simple febrile seizures, while balance 8 children had complex seizures. Among the latter, two cases met the criteria for febrile status epilepticus, with seizures lasting over 30 minutes. First-time seizure episodes were more common (64%) compared to recurrent seizures (36%).

These demographic and clinical characteristics are summarized in Table 1.

TABLE 1 Demographics and clinical presentation of febrile seizures

Variable	Category	Number (n)	Percentage (%)
Age Group	<1 year	6	24.0
	1-2 years	9	36.0
	2-5 years	9	36.0
	>5 years	1	4.0
Mean Age (SD)	-	2.63 years	(±2.22 years)
Gender	Male	14	56.0
	Female	11	44.0
Seizure Type	Simple	17	68.0
	Complex	8	32.0
Episode	First Presentation	16	64.0
	Recurrent	9	36.0

Upper respiratory tract infections (URTI) were the most frequently identified cause of fever in 14 (56%), followed by viral fever (20%) and gastroenteritis (8%). One episode occurred following OPV and DTP (4th dose) vaccination. In 12% of cases, no clear cause was identified. Complications were recorded in two children: one developed aspiration pneumonia, while another showed transient unilateral weakness post-seizure.

Table 2 summarizes the causes of fever and the investigations performed according to seizure type.

In terms of investigations, all children with complex febrile seizures were investigated while only 65% of children with simple febrile seizures had blood or urine examined. Notably, none of the children were subjected to lumbar puncture, EEG, or neuroimaging (Table 2).

For ongoing seizures (5 cases), oxygen was administered, and all received basic life support in line with APLS guidance. Seizure termination was attempted with midazolam as the first-line antiepileptic in all five cases. Of these, 40% received it intravenously, and 60% via the buccal route. Due to the absence of buccal midazolam preparations, the intravenous formulation was repurposed and administered buccally using a syringe without a needle at a dose of 0.3 mg/kg.

Paracetamol was administered in all patients (100%) as the sole antipyretic. Doses were appropriately calculated based on documented weight, and mothers were educated to avoid double dosing in all cases. Paracetamol suppositories were used in 63% of cases (7 out of 11), and oral syrup in 36% (4 out of 11). Temperature was documented prior to administration in all cases. While 40% had undergone tepid sponging and 24% had been fanned by caregivers before arrival, none of these methods were encouraged in the PCU.

Following seizure resolution, the most common reason for admission to the ward was first-time seizure presentation (14 out of 21 admissions). Complex febrile seizures accounted for 14.3% (3 out of 21), and parental concern for 9.5% (2 out of 21). However, two children who experienced their first simple febrile seizure were discharged without admission, contrary to standard recommendations. For recurrent simple febrile seizures, observation in the PCU for six hours was implemented in 4 out of 9 cases, as per protocol.

A significant finding of the audit was the inadequacy of parental education. Only 32% of cases (8 out of 25) received verbal instructions on febrile seizure management, while written advice was documented in merely 16% (4 out of 25). Information delivery was inconsistent, lacking structure and uniform timing. Among those with recurrent seizures, only 22% (2 out of 9) received any counseling, compared to 50% (8 out of 16) of first-time cases, (Table 3). Eight key points identified as essential in febrile seizure education were assessed during caregiver interactions. While some aspects, such as seizure safety practices and recurrence risks, were mentioned in 50-75% of cases, others, especially the reassurance that febrile seizures do not significantly increase the risk of epilepsy, were never discussed.

Intermittent oral clobazam was prescribed in one case (4%), although there is no evidence supporting its use as a prophylactic medication. No prehospital use of buccal or nasal antiepileptics by caregivers or paramedics was noted in any case.

TABLE 2 **Investigations performed according to seizure type**

Seizure Type	Total Cases (n) Cases, n (%)	Investigated	Common Investigations	No Investigations, n (%)
Simple Febrile Seizure	17	11 (65%)	CBS (11), FBC (4), VBG (1), UFR (1), Urine ketones (1)	6 (35%)
Complex Febrile Seizure	8	8 (100%)	CBS (6), FBC (5), VBG (3)	0 (0%)

Abbreviations: CBS – Capillary Blood Sugar; FBC – Full Blood Count; VBG – Venous Blood Gas; UFR – Urine Full Report

TABLE 3 Overview of parental education on febrile seizures

Education Type	Number (n)	Percentage (%)
Overall Cases (n=25)		
Verbal Education Provided	8	32.0
Written Education Provided	4	16.0
Subgroup Analysis		
Education in First Febrile Seizure Cases (n=16)	8	50.0
Education in Recurrent Seizure Cases (n=9)	2	22.2
Content of Education Among Counseled Cases (N = 8)		
What is a febrile seizure and how to recognize	8	100
Common and benign condition	8	100
Not due to CNS infection	4	50
No increased epilepsy risk	0	0
Age-dependent, child outgrows it	4	50
Risk factors for recurrence	5	62.5
What to do during future seizures	6	75
What not to do during seizures	5	62.5

DISCUSSION

The demographic distribution of febrile seizures in our audit aligns with established literature, showing slight male predominance,[3] and a peak incidence between 12 and 18 months.[4] The observed ratio of simple to complex febrile seizures (72% vs 28%) reflects global trends.[5]

URTIs emerged as the leading cause of febrile seizures,[6] consistent with international findings suggesting viruses, particularly those with neurotropic properties,[7] are often involved. The single vaccine-associated case is likely due to the fever-inducing potential of the whole-cell pertussis component in DTP,[8] reinforcing the importance of addressing vaccine-related parental fears.[9]

Over-investigation was noted in simple febrile seizures, despite guidelines discouraging routine labs, EEG, or imaging unless atypical features exist.[10] This likely reflects cautious but unnecessary practice.[11] In contrast, investigations in complex seizures were appropriate to exclude metabolic or infectious causes.[12] Avoiding lumbar puncture, EEG, and imaging in the absence of red flags was consistent with guidelines.

Paracetamol use in the PCU was appropriate in terms of dosage and timing for fever management and child comfort.[13] While intermittent antipyretic use is justified, routine regular use in all febrile children remains controversial,[14] as evidence shows it does not reduce seizure recurrence.[15] Additionally, non-conventional methods like tepid sponging and fanning were not encouraged in the PCU, reflecting evidence-based practice.[16]

Use of midazolam adhered to APLS guidelines[17] Current unit practice prioritizes IV midazolam; IM is used if IV access is unavailable, with buccal as a last resort. Though the off-label buccal administration of IV midazolam presents concerns. While effective and resourceful, such practices carry risks, including mucosal irritation, dosing errors, and poor palatability, and underscore the need for proper drug formulations.[18]

Under-admission was observed in the unit, deviating from guidelines, as first simple febrile seizures require admission to exclude causes like CNS infections or metabolic disorders, failure to do so may compromise diagnostic safety. The observation in the PCU for several hours after the seizure probably was the reason since this facilitates observation

for possible features of CNS infection. However, observation periods for recurrent seizures adhered to guidelines.

Parental education remains a critical shortcoming, with crucial information often omitted. One important area relates to misconception of febrile seizures developing into epilepsy, believed by 65% of parents in Sri Lanka.[19] Failure to address these misconceptions may contribute to parental anxiety. A study at LRH found no link between prior febrile seizures and parental knowledge, supporting the need for re-education at every visit including during recurrences.[20] PCU had a pre-formed leaflet, but distribution was limited by the country's economy.

The single observed case of intermittent clobazam use is reassuring and suggests overall guideline adherence.[14] However, it still reflects residual non-recommended practice, likely driven by caregiver anxiety, limited rural healthcare access, and low parental confidence.

RECOMMENDATIONS

To improve the quality of care, over-investigation and under-admission should be addressed through implementation of a standardized PCU protocol. Ensuring availability of appropriate formulations, especially buccal midazolam, would enhance safety and compliance. Parental education should be standardized using structured protocols and multilingual materials, supported by routine caregiver feedback, to enhance understanding and confidence. Ongoing staff training and periodic re-auditing will further reinforce adherence to guidelines and foster sustainable quality improvements.

CONCLUSION

While the PCU at Lady Ridgeway Hospital demonstrates overall good performance in clinical management of febrile seizures, several inconsistencies were identified. With targeted interventions and structured re-evaluation, the unit can elevate its standard of care related to management of febrile seizures.

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SLJoN

ORIGINAL RESEARCH

Non-traumatic spinal cord injuries: A descriptive study at the National Hospital of Sri Lanka

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Abstract

Background: Non traumatic spinal cord injury (NTSCI) is becoming increasingly prevalent worldwide, particularly with the rise in ageing populations. Despite this global trend, there is a lack of comprehensive data on the epidemiology and aetiology of NTSCI in Sri Lanka. This study aims to provide an epidemiological overview of NTSCI cases in a Sri Lankan cohort, focusing on demographic characteristics, clinical presentations, aetiologies, and outcomes.

Methods: This descriptive study included all patients diagnosed with NTSCI who were admitted to the neurology wards or attended neurology clinics at the National Hospital of Sri Lanka (NHSL). Patients were selected based on predefined inclusion and exclusion criteria. Data were collected using structured clinical records and analyzed using SPSS software. Variables assessed included demographic details, clinical features, investigation findings, and aetiological diagnoses.

Results: The study population had a mean age of 48.17 years (range: 17-78 years). Paraparesis was the most common presentation, reported in 81.5% of patients, while 18.5% presented with tetraplegia. A sensory level was documented in 42.6% of cases. Bladder and bowel involvement were observed in 48.1% of patients. Regarding aetiology, myelitis and demyelinating conditions accounted for the largest proportion (42.6%), followed by degenerative disc disease and disc prolapse (35.2%). Rare causes included syringomyelia and hereditary disorders (1.9%). A significant majority (93.6%) of patients underwent and responded positively to rehabilitation programs.

Conclusion: NTSCI in Sri Lanka primarily affects individuals in their late 40s, with both males and females represented. The most frequent aetiologies are demyelination, myelitis, and degenerative spinal conditions. Sensory, motor, and autonomic dysfunctions are common. Rehabilitation outcomes are generally favourable. These findings highlight the need for improved surveillance, early diagnosis, and targeted rehabilitation services to optimise care for NTSCI patients in Sri Lanka.

KEYWORDS

Non traumatic spinal cord injury, aetiology, myelitis, demyelination, degenerative disc disease

INTRODUCTION

Spinal cord injury (SCI) can result in motor, sensory or autonomic dysfunction, often leading to significant disability and reduced quality of life. The damage to the spinal cord arising from non- traumatic causes is referred to as non-traumatic spinal cord injury (NTSCI).[1]

An Australian study reports the average age-adjusted incidence rate of NTSCI in adults as 26.3 cases per million per year.[2] The global incidence of spinal cord injuries (primarily traumatic) is estimated at 15 to 40 cases per million.[3] There is significantly less published data on NTSCI compared to TSCI. A study from the Republic of Ireland done in 2017 reported an incidence of 26.9 per million per year.[4] There is a trend of increasing incidence and prevalence of NTSCI with advanced age.[2] An Australian study projected the prevalence of NTSCI in adults over 16 years to increase from 367.2 to 455 per million by the year 2027.[5] NTSCI is more common in males and is extremely rare in children.[2] Traumatic spinal cord injury (TSCI) is more prevalent among young adults, posing a substantial burden on affected individuals, families and society.[3]

NTSCI can have different aetiologies.[1] In the West, the common causes of NTSCI include degenerative disc disease, spinal canal stenosis, tumours, vascular disorders and inflammatory conditions.[6,7] There is lack of data on the aetiology and epidemiology of NTSCI in Sri Lanka.

According to global data, there is a rising trend of NTSCI with ageing of the population.[1] Therefore, assessing the incidence, prevalence, aetiology and survival following NTSCI is crucial due to this rising trend and the social burden. This is essential in order to plan health care services for the current demand and aim for the future burden. The aim of this study is to assess the situation in a selected group of NTSCI patients in Sri Lanka.

METHODS

The study was designed as a prospective descriptive study and was conducted at the National Hospital of Sri Lanka (NHSL) from 01.04.2021 to 31.12.2021 over a nine-month period. All patients with NTSCI admitted to the neurology wards or attending neurology clinics at NHSL were considered for inclusion, based on pre-defined eligibility criteria. All patients presenting with clinically evident paraparesis and tetraparesis with or without sensory symptoms with a compatible spinal cord lesion on spinal magnetic resonance imaging (MRI) were included. Exclusion criteria included patients with traumatic spinal cord injuries, those under 16 years of age or individuals with neurological symptoms due to perinatal complications or cerebral palsy. Data collection was continued until the minimum sample size was reached. Consented participants were interviewed with an interviewer administered questionnaire. A complete examination of the nervous system was carried out by one of the investigators and the patient's records and imaging studies were reviewed and evaluated.

RESULTS

A total of 54 patients were included in the study. The mean age of the study population was 48.17 years (standard deviation (SD) 16.16) with ages ranging from 17 to 78 years. Of the participants 28 were males (51.9%) and 26 were females (48.1%). The majority 59.3% (32/54) were from the Western Province and 77.8% (42/54) were married. Only 31.5% (17/54) were employed at the time of data collection.

Paraparesis was the most common presentation, occurring in 81.5% (44/54) of patients, with 18.5% (10/54) having tetraplegia. Most patients had an incomplete SCI according to the American Spinal Cord Association (ASIA) Impairment scale (Table 1). Among the patients, 3 had a tetraplegic-complete SCI, 7 had tetraplegic-incomplete, 17 were paraplegic complete, and 27 were paraplegic-incomplete. Within the study population, 42.6% (23/54) had a defined sensory level, 48.1 % (26/54) had bladder involvement and 48.1 % (26/54) had bowel involvement.

Imaging studies revealed that 61.1% (33/54) of patients demonstrated spinal cord hyperintensity on MRI and 35.2% (19 /54) had degenerative changes including disc prolapse and root compression. Dorsal and lumbosacral spinal segments were involved in 50% (27/54) of the cases.

The aetiology of NTSCI of the study population is presented in Table 2.

TABLE 1 Presentation according to the ASIA Impairment scale

Presentation	A	B	C	D	E
Paraparesis	17	18	7	2	0
Tetraparesis	3	5	2	0	0

TABLE 2 Aetiology of NTSCI

Aetiology	Number	Percentage
Disc prolapse Degenerative spine	19	35.2
Myelitis, Demyelination	23	42.6
Tumour, Metastasis	2	3.7
Tuberculosis/ Melioidosis	3	5.6
Metabolic, nutrient deficiency	5	9.3
Syrinx	1	1.9
Hereditary	1	1.9

Based on aetiology and age, the age range for degenerative spinal disease and disc prolapse leading to NTSCI was between 46 and 78 years, encompassing 37% patients in our study (20/54). For demyelinating diseases, the age range was broader, spanning from 17 to 60 years, and involved 23 (42%) of patients. Patients presenting with syringomyelia and hereditary causes were aged 28 and 38 years, respectively. Metabolic and infective aetiologies were observed in patients aged between 20 and 68 years. Tumour related spinal cord presentations occurred at ages 42 and 57.

Rehabilitation outcomes were favourable, with 93.6% (52/54) of patients showing satisfactory follow-up results. Rehabilitation protocols were initiated with inpatient physiotherapy. Patients with significant neurological compromise were transitioned to institutional rehabilitation programs, whereas those with less severe impairment continued with outpatient-based rehabilitation.

DISCUSSION

To our knowledge, this is the first study to investigate the epidemiology of non-traumatic spinal cord injury (NTSCI) among adults in Sri Lanka. In our study, NTSCI was more common in males (51.9%) than females (48.1%), consistent with trends in the world literature. In our population, the mean age of NTSCI was 48.17 years which is the common age group described in developed countries as well.

In terms of aetiology, demyelinating diseases and myelitis were the most prevalent causes of NTSCI in our study, though degenerative spine disease and spinal tumours were more frequently reported in the international studies.[6] Spinal tumours were relatively less in our population. Syringomyelia and hereditary disorders were rare, reflecting their overall low prevalence.

The study demonstrates a notable pattern in aetiology relative to age distribution. Degenerative spinal pathologies were predominantly observed in the older population, whereas demyelinating diseases were distributed across a wider age spectrum, with a significant representation among middle-aged and younger individuals.

We found that incomplete paraplegia was the most common injury reported in our study which is similar to the study done in Ireland.[5] It was reported that paraplegia was more common than tetraplegia in the Australian and Canadian studies where complete or incomplete status was not reported.[6] Rehabilitation outcomes were favourable, with 93.6% of patients showing satisfactory follow-up results.

CONCLUSION

The mean age of patients with non-traumatic spinal cord injury (NTSCI) in Sri Lanka was 48. The most common aetiologies were demyelinating diseases, myelitis and degenerative spinal conditions. A high rate of rehabilitation follow-up was observed, which may help mitigate the long-term disability burden associated with NTSCI in the community.

Conflicts of interest

There are no conflicts of interest associated with the publication and the authors received no financial support for the research, authorship and/or publication of this article.

Author contributions

N.W. conceived the study. A.F. developed the methodology. N.W., P.M., S.B., and A.F. were involved in case examination and data collection. N.W. performed the statistical analysis and drafted the final manuscript. A.F. and S.B. provided supervision. All authors reviewed and approved the final version of the manuscript and its revision.

N.W. conceived the study. A.F. developed the methodology. N.W., P.M., S.B., and A.F. were involved in case examination and data collection. N.W. performed the statistical analysis and drafted the final manuscript. A.F. and S.B. provided supervision. All authors reviewed and approved the final version of the manuscript and its revision.

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Ethical clearance

This study received ethical clearance from the Ethics Review Committee of the National Hospital of Sri Lanka (NHSL), in accordance with institutional guidelines and ethical standards.

Data accessibility statement

Due to patient confidentiality, data are not publicly available, but may be obtained from the corresponding author upon reasonable request.

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ORIGINAL RESEARCH

The epidemiology of stroke in Northern Sri Lanka: A population-based descriptive cross-sectional study

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Abstract

Introduction: Stroke is a significant health concern in Sri Lanka, leading to high rates of mortality and morbidity. Healthcare must prioritize prevention and early detection to address this issue. While research on stroke treatment in Sri Lanka is limited, local studies highlight changing trends and risk factors. This study aimed to assess the prevalence of stroke and its risk factors in the Northern Province of Sri Lanka.

Methodology: This population-based cross-sectional study conducted by the NIHR Global Health Research Group on Atrial Fibrillation focused on individuals aged 50 and above in the Northern Province. Using a multi-stage sampling approach, the study recruited a sample size of 10,000 Tamil-speaking participants representing all five northern districts. Data were collected with an interviewer-administered questionnaire. Descriptive statistics was used to determine the prevalence and risk factors.

Results: Out of 10,000 individuals, 231 had a stroke, resulting in a 2.3% prevalence rate. Among the 231 patients with stroke, mean age was 68.6 (SD 8.6) years, with 54.1% and 45.9% being males and females, respectively. Educational status varied, with most having primary education or less (40.2%). Retirees comprised 47.6% of the group. Common risk factors included hypertension (71.0%), diabetes mellitus (34.6%) and palpitations (30.7%).

Conclusion: Our study revealed a higher stroke prevalence rate (2.3%) in the Northern Province compared to the national rate (1.0%). Patients with stroke had a high prevalence of risk factors such as hypertension, diabetes and heart diseases. These insights highlight the need for tailored primary prevention and management strategies, considering socio-economic factors and specific regional risks, particularly screening programs and rehabilitation services.

KEYWORDS

Stroke, Sri Lanka, elders, risk factors, Northern Province

INTRODUCTION

Stroke is a major health issue in Sri Lanka, with a significant effect on death and illness rates. It is a the fifth highest cause of death in Sri Lanka according to Ministry of Health statistics.[1] The impact of stroke goes beyond just deaths – it also results in decreased work capacity and notably raises the expenses related to hospitalization. Additionally, the economic impact of stroke places considerable strain on families and society overall. It is therefore crucial for the healthcare system to prioritize preventive actions and early detection to manage this problem effectively.

While local studies on the epidemiology of stroke are scarce, recent research highlights the increasing prevalence of stroke among young adults in Sri Lanka, shedding light on

the changing demographics of stroke occurrence in the country.[2] Another study that explored the association between dietary habits and stroke risk in the Asian region, including in Sri Lanka, revealed changes in the contribution of unhealthy diet to the disease burden of stroke by time and place.[3] The 2021 Risk Factor Survey conducted by the Ministry of Health revealed that the prevalence of several risk factors for stroke, including obesity, physical inactivity, alcohol consumption, hypertension, had increased between 2015 and 2021.[4] Moreover, research on the genetic susceptibility to stroke has contributed significantly to understanding the hereditary factors influencing stroke in the Sri Lankan population.[5]

The Northern Province of Sri Lanka is located just 22 miles (35km) southeast of India and has approximately 1.3 million permanent residents. It is made up of five districts: Jaffna, Kilinochchi, Mannar, Mullaitivu and Vavuniya. Each district is divided into administrative units called Divisional Secretariats (DS). Each DS is further divided into many Grama Niladhari (GN) divisions. Publicly available healthcare, including medical appointments, medications and medical procedures, are free to all Sri Lankan citizens, including in the Northern Province.

Little is known about the epidemiology of stroke in northern Sri Lanka. Considering the global burden of stroke, the rising prevalence of risk factors and the healthcare system challenges faced by the country, it is imperative to conduct research on the epidemiology of stroke. Therefore, this study aimed to assess the prevalence of stoke and its risk factors in the Northern Province of Sri Lanka.

METHODS

Data were obtained from the 'Prevalence of atrial fibrillation (AF) in Northern Sri Lanka' study carried out by the NIHR Global Health Research Group on Atrial Fibrillation.[6] This cross-sectional study commenced in June 2020 and was completed in March 2022. Ethics approval was attained from the Ethics Review Committee of the Faculty of Medicine, University of Jaffna.

The study was conducted across all five districts in the Northern Province. In line with similar studies conducted in lower-middle-income countries and considering the growing body of literature on disease prevalence in South Asia, we set the screening inclusion age at 50 years and above, which is below the current recommended practice of 65 years. Therefore, individuals aged 50 years or older who are proficient in Tamil were eligible for screening. As per established methods published elsewhere, individuals with terminal illnesses, those requiring immediate hospitalization, or currently admitted as hospital inpatients, were not included. The team of data collectors comprised a qualified medical doctor and two nursing graduates.

This research employed a sampling methodology that involves multiple stages. The process commenced at the district level, then proceeded to the DS level, and finally reached the GN level. Subsequently, GN divisions were grouped into clusters based on population size, from which one cluster was chosen randomly. Each cluster encompassed 20 households with one participant per household. Initially, an index house within each cluster was selected randomly followed by picking 20 households located on its right side. In cases where there were multiple eligible individuals in a household, the individual whose birthday is closest to the date of visitation was chosen. Census data from 2012 were utilized for selecting both the clusters and index houses within each cluster.

Based on previous evidence, the prevalence of AF in Sri Lanka was estimated to be 1%. A design effect of 2 was applied to account for cluster sampling, with an alpha level set at 5% and a beta level at 20%. To accommodate non-participation, the sample size was increased by 10%, resulting in a minimum requirement of 10,000 participants. Previously conducted research has indicated that the prevalence of stroke is also approximately 1%.[7] Therefore, this sample size was deemed suitable for determining the occurrence rate of stroke in the Northern Province. Descriptive analysis was performed to identify the prevalence of the stroke and AF.

RESULTS

Out of a total sample size of 10,000 individuals, 231 had experienced a stroke, resulting in a prevalence rate of 2.3%. Prevalence of stroke varied by district (Table 1). The highest prevalence was in the Mannar district (3.1%) and lowest in Kilinochchi district (1.8%) but this variation was not significant (X^2=3.2, df=4, p=0.5).

TABLE 1 Prevalence of stroke across the Northern Province (n=231)

District	n (%)
Mannar	26 (3.1%)
Jaffna	144 (2.3%)
Vavuniya	31 (2.2%)
Mullaitivu	14 (2.1%)
Kilinochchi	16 (1.8%)

The average age was 68.6 (SD ±8.6) years. In terms of gender distribution, there were 125 males (54.1%) and 106 females (45.9%). Among participants, 135 (58.4%) were living with their partner. Evaluation of educational status revealed that 87 (37.7%) had only primary education with 6 (2.6%) having no formal education at all. In the sample, 110 (47.62%) were retired, 91(39.39%) were homemakers, while 23(9.95%) were employed full time (Table 2).

TABLE 2 Socio-demographic characteristics of stroke patients (n=231)

	n (%)
Gender	
Male	125 (54.1)
Female	106 (45.9)
Marital status	
Living with partner	135 (58.4)
Not living with partner	96 (41.6)
Educational level	
University degree and above	5 (2.3)
Diploma	3 (1.3)
GCE A/L	10 (4.3)
GCE O/L	43 (18.6)
Middle school	77 (33.3)
Primary and less	93 (40.2)
Occupation	
Employed	23 (9.9)
Retired	110 (47.6)
Homemakers	92 (39.3)
Other	7 (3.2)

Of the risk factors, 164 (71.0%) had hypertension, 80 (34.6%) had diabetes mellitus, 71 (30.7%) had palpitations, 30 (13.0%) had chronic kidney disease, and 43 (18.6%) had ischaemic heart disease (Table 3).

TABLE 3 Risk factors of stroke (n=231)

Variables	n (%)
Symptoms	
Palpitations	71 (30.7)
Comorbidities	
Hypertension	164 (71.0)
Diabetes	80 (34.6)
Ischemic heart disease	43 (18.6)
Chronic kidney disease	30 (13.0)
Valvular heart disease	9 (3.9)
Heart failure	1 (0.4)

DISCUSSION

The results of our study in the Northern Province of Sri Lanka have provided significant insights into the prevalence and risk factors associated with stroke in this specific region. The prevalence of stroke was found to be notably higher at 2.3% compared to the 1% occurrence rate observed in other national research studies.[8] A study done in India also showed a prevalence of 1.5% .[8] This disparity could be attributed to regional variations in stroke occurrence, or it may be a result of the difference in methodology and the substantial sample size employed in our study. Further studies are needed to understand the unique characteristics and challenges associated with stroke in the Northern Province to guide the development of more precise and impactful prevention strategies.

One significant aspect is to consider is the distribution of risk factors in the subsample of patients with stroke. A substantial portion of the participants had hypertension, with 71% of the sample affected. Additionally, the prevalence of diabetes mellitus, at 34.6%, and the presence of ischemic heart disease, at 18.6%, further emphasize the need for strengthening screening programmes to identify these risk factors early. It is worrying that recent risk factor surveys reveal a rising prevalence of obesity, physical inactivity, hypertension, and other risk factors of stroke.[4]

Furthermore, the average age of participants who experienced a stroke was found to be 68.6 years. This finding highlights the vulnerability of the elderly population to stroke in the Northern Province and emphasizes the importance of age as a risk factor.[9] The educational status of those with stroke also reveal interesting patterns, with a substantial proportion having only received primary education or lower, indicating the potential influence of socio-economic factors on stroke risk.[10]

These findings underscore the importance of tailored prevention and management strategies for stroke in the Northern Province. It is evident that interventions focusing on controlling hypertension, managing diabetes, and addressing cardiovascular risk factors can play a crucial role in reducing the burden of stroke in this region. Integrating these insights into healthcare policies and clinical practice can lead to more effective interventions and better outcomes for the population of the Northern Province. In addition, healthy public policies are urgently needed to control the exposure to risk factors, especially among young people.[11]

While our study found a high prevalence of hypertension, diabetes mellitus, and cardiovascular conditions in patients experiencing a stroke, future research could focus on

evaluating the effectiveness of tailored prevention and management strategies targeting these risk factors specifically within the context of the Northern Province. This could involve conducting longitudinal studies to track the impact of targeted interventions on the prevalence of stroke in the region, as well as assessing the feasibility and acceptability of these strategies within the local healthcare infrastructure.

Another important aspect that warrants further investigation is the influence of socio-economic factors on stroke risk within the Northern Province. Exploring the socioeconomic determinants of health, such as income level, access to education, and employment status, could provide valuable insights into the underlying social disparities that contribute to the burden of stroke in this region. Conducting epidemiological studies that integrate socio-economic indicators into the analysis of stroke prevalence and risk factors could inform the development of more equitable and inclusive public health interventions.

CONCLUSION

This study revealed a higher stroke prevalence rate (2.3%) in the Northern Province compared to the national rate (1.0%). Patients with stroke had a high prevalence of risk factors such as hypertension, diabetes and heart diseases. These insights highlight the need for tailored primary prevention and management strategies, considering socio-economic factors and specific regional risks, particularly screening programs and rehabilitation services.

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Conflict of interests

The authors have no conflict of interests to declare.

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CASE REPORT

Severe neurological manifestations of LETM with brainstem involvement: A dual case report

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Abstract

Background: Transverse myelitis (TM) is a rare neurological disorder characterized by inflammation of the spinal cord, which can cause varying degrees of motor, sensory, and autonomic dysfunction. Longitudinally extensive transverse myelitis (LETM) refers to TM that extends over three or more vertebral segments and may be associated with brainstem involvement, leading to complex clinical symptoms.

Case presentation: This case report presents two cases of LETM with concurrent brainstem involvement, both the patients exhibited progressive neurological deficits, including limb weakness, sensory disturbances and cranial nerve related symptoms.

Imaging findings and management: MRI examination of the spine revealed T2 Hyperintense lesions extending over multiple vertebral segments, consistent with LETM. Brain MRI in both cases showed hyperintensities involving the medulla and pons, indicating brainstem involvement. The patients were managed with high-dose corticosteroids and supportive therapy. One of the patients showed marked clinical improvements, while the other required prolonged rehabilitation with partial neurological recovery.

Conclusion: LETM with brainstem involvement represents a severe neurological condition with complex clinical and radiological features. Early diagnosis through MRI and prompt immunosuppressive therapy are essential for improved outcomes.

KEYWORDS
Longitudinally extensive transverse myelitis (LETM), MRI features in LETM, Brainstem involvement in LETM

INTRODUCTION

Myelopathy is any form of spinal cord pathology whereas myelitis refers to an inflammatory or infectious process.[1] Transverse myelitis (TM) is an infectious or inflammatory disorder that affects the spinal cord and typically disrupts the transmission of nerve signals, leading to symptoms such as paralysis, sensory loss, pain, and autonomic dysfunction. When the inflammation spans more than three segments of the spinal cord, it is classified as longitudinally extensive transverse myelitis (LETM).[2] LETM is often associated with more severe outcomes and may have an underlying etiology, including infections, autoimmune diseases, or demyelinating conditions like multiple sclerosis (MS). In rare cases, brainstem involvement may occur, complicating the presentation as well as management. The radiological features in MRI can suggest or help to differentiate the various etiologies of LETM.[3,4]

CASE PRESENTATION

Case 1

A 32-year-old male presented to the emergency department with complaints of rapidly progressive weakness and numbness in both legs, which had developed over the past 72 hours. He also reported dizziness, dysphagia, and blurred vision. The patient had no significant past medical history but had a recent viral upper respiratory infection. On neurological examination, the patient exhibited paraplegia, dysarthria, and dysphagia. MRI of the cervical and thoracic spine was done which showed presence of longitudinally extensive lesions spanning from C2 to T1 and D3 to D7, characterized by hyperintensity on T2-weighted images and hypo-intensity on T1-weighted images. These lesions appeared to involve both gray and white matter of the spinal cord, with no evidence of significant atrophy or structural abnormalities of the surrounding vertebrae. The MRI of the Brain revealed subtle hyperintensities on T2-weighted images in the brainstem, especially in the pons on right side. There was no gadolinium enhancement in the area. These findings were consistent with brainstem involvement in the context of transverse myelitis. There was no significant lesions were noted in other parts of the brain, which helped exclude diagnoses such as multiple sclerosis or other demyelinating diseases. The inference drawn from these findings was that of LETM with brainstem involvement, secondary to an infectious etiology (Figure1).

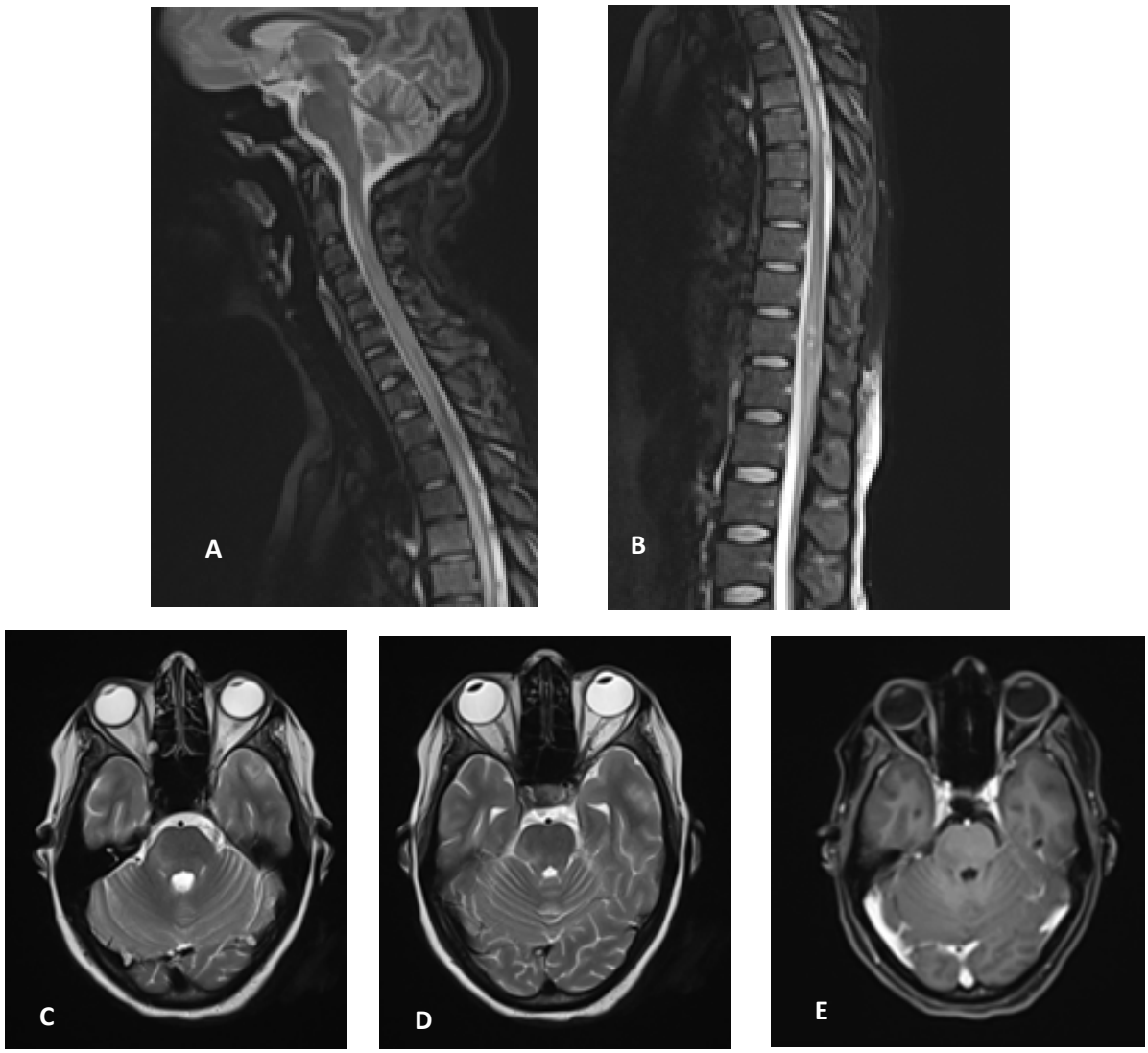

FIGURE 1 **MRI of the cervical and thoracic spine (A and B, STIR sagittal cervical and thoracic, C and D axial T2W, E post contrast axial) showed presence of longitudinally extensive lesions spanning from C2 to T1 and D3 to D7, characterized by hyperintensity on T2-weighted images and hypo-intensity on T1-weighted images without atrophy of the cord. The MRI of the Brain revealed subtle hyperintensities on T2-weighted images in the brainstem, especially in the pons on right side. There was no gadolinium enhancement in the area.**

Case 2

A 42-year-old female presented with acute onset of weakness and sensory loss in both lower limbs, accompanied by bladder dysfunction and loss of deep tendon reflexes. There was no history of trauma, and the patient was previously healthy. The symptoms developed rapidly over the course of 48 hours. Neurological examination revealed signs consistent with myelopathy, including impaired motor strength, decreased sensation below the T10 dermatome, and absent reflexes. The MRI of the spine in this case revealed a longitudinally extensive lesion involving the cervical and thoracic spinal cord, spanning from C3 to T10 with mild expansion of the cord. The lesion appeared hyperintense on T2-weighted images and hypointense on T1-weighted images. The MRI of Brain in this case also showed a lesion in the brainstem, predominantly involving the pons (Left>right), with areas of hyperintensity on T2-weighted images. No significant gadolinium enhancement was observed in the brainstem lesions. The conclusion drawn in this case was also LETM with brainstem involvement – suspected to be of demyelinating etiology (Figure 2).

The patients were managed with high-dose corticosteroids and supportive therapy. the first case patient showed marked clinical improvements, while the second case required prolonged rehabilitation with partial neurological recovery.

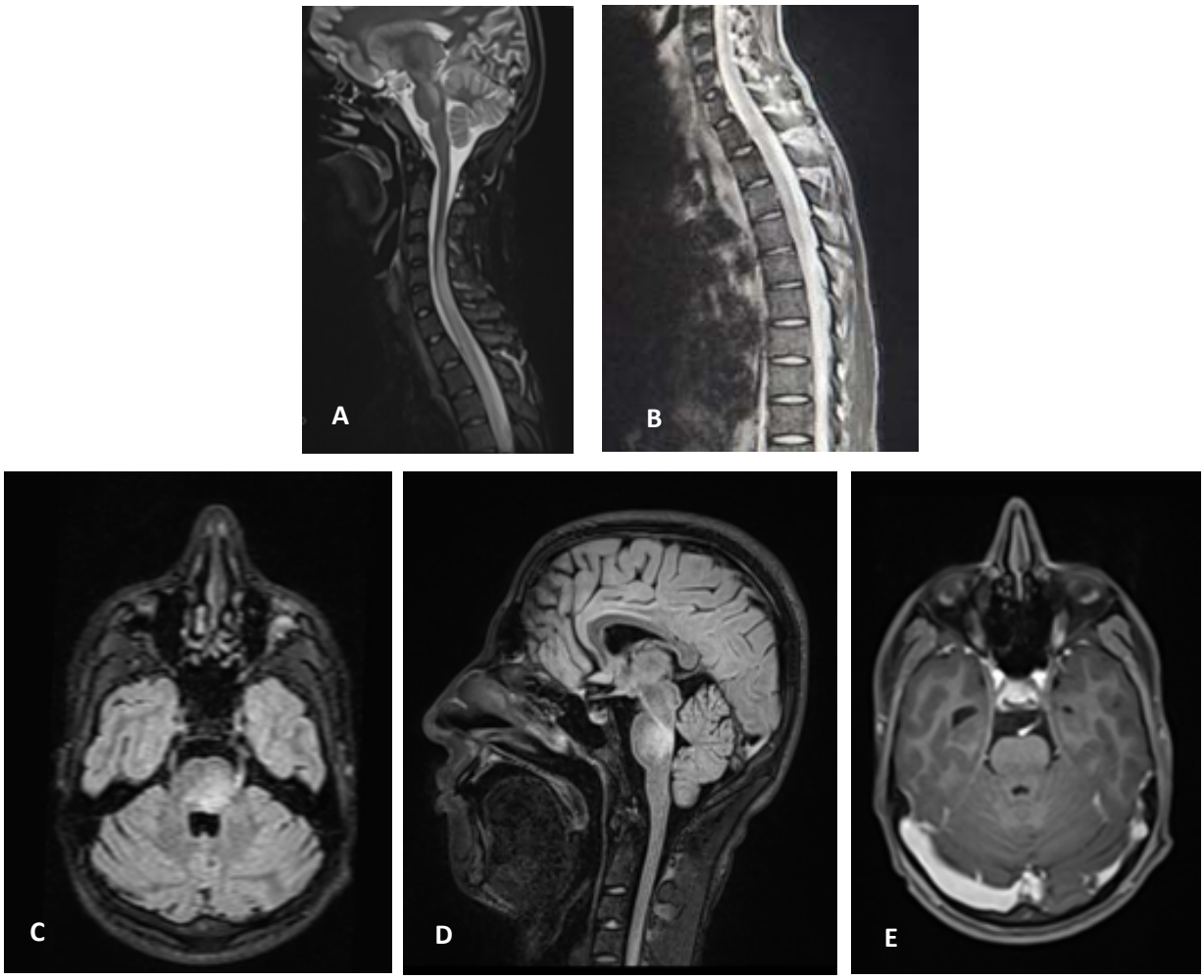

FIGURE 2 The MRI of the spine (A- sagittal STIR cervical , B-Sagittal T2W Thoracic, C and D axial and sagittal FLAIR brain and D-Axial brain post contrast) revealed a longitudinally extensive lesion involving the cervical and thoracic spinal cord, spanning from C3 to T10 with mild expansion of the cord. The lesion appeared hyperintense on T2-weighted images and hypointense on T1-weighted images. The MRI of Brain in this case also showed a lesion in the brainstem, predominantly involving the pons(Left>right), with areas of hyperintensity on T2-weighted images. No significant gadolinium enhancement was observed in the brainstem lesions.

DISCUSSION

Longitudinally extensive transverse myelitis has characteristics of contiguous lesions in the spinal cord and when brainstem involvement presents a diagnostic challenge due to the wide array of differential diagnoses, viral infections, and autoimmune disease neuromyelitis Optica and rarely with multiple sclerosis.[3,4] The clinical presentation is usually with single or multiple attacks of paraparesis or tetra-paresis, sensory deficits and bladder/bowel disturbances and may even progress to respiratory failure.[5] The brainstem involvement, although less common in LETM, may explain the more severe clinical manifestations, including cranial nerve dysfunction, dysphagia, and facial weakness. The detection of such lesions on brain MRI is crucial for accurate diagnosis and management, especially when a concomitant inflammatory or autoimmune disorder is suspected.[7] The MRI findings in our case series reveal characteristic patterns of demyelination and infection in both the spinal cord and brainstem.

Additionally, longitudinal involvement of the spinal cord and brainstem on MRI may help differentiate LETM from other spinal cord pathologies like multiple sclerosis, which typically involves discrete lesions separated by normal-appearing tissue.[8-10]

The following is to be considered in this clinical and radiological settings:

- **Multiple Sclerosis (MS):** Characterized by multiple demyelinating plaques in the central nervous system (CNS). However, MS typically involves fewer than three spinal segments and may show more discrete brain lesions.
- **Neuromyelitis Optica Spectrum Disorder (NMOSD):** This condition is associated with optic neuritis and transverse myelitis but often presents with optic nerve and spinal cord involvement rather than brainstem involvement.
- **Viral Infections:** Such as herpes simplex virus (HSV) or Epstein-Barr virus (EBV) can cause transverse myelitis with or without brainstem involvement, but the rapid progression of symptoms was not typical for viral etiology.
- **Autoimmune Disorders:** Such as systemic lupus erythematosus (SLE) or vasculitis, can cause myelitis but were excluded by the absence of systemic signs of active disease.

  The presence of LETM mandates a prompt search for disease entities like neuromyelitis Optica and myelin oligodendrocyte glycoprotein.

CONCLUSION

LETM with brainstem involvement is a complex neurological disorder requiring high clinical suspicion and advanced imaging techniques for accurate diagnosis. MRI remains the cornerstone of diagnosis, and in cases with brainstem involvement, it is essential for distinguishing between potential etiologies. The radiological findings in this case series underscore the importance of early detection and monitoring of these lesions to guide treatment decisions and improve patient outcomes. Further studies on the long-term prognosis and treatment responses in patients with LETM and brainstem involvement are needed.

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Neuropalliative care

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Abstract

"Five Key Practice Aspects" is a thoughtfully curated series of articles authored by expert neurologists to distill and highlight the most critical facets of everyday clinical practice within various neurology subspecialties. Each article in this series identifies and elaborates on the five most essential considerations in the diagnosis and management of neurological conditions, combining robust evidence-based insights with the seasoned clinical experience of specialists in the field.

The goal of this series is to serve as a concise yet comprehensive reference that caters to both the trainee seeking foundational knowledge and the seasoned neurologist looking for a practical refresher. By focusing on the aspects that are most relevant and actionable in real-world practice, this series offers a streamlined guide that supports informed decision-making across diverse clinical scenarios.

Designed for ease of access and utility, "Five Key Practice Aspects" is a portable resource for busy clinicians who need reliable information on the go. Whether navigating complex cases or honing subspecialty skills, this series aspires to become an indispensable companion in the pursuit of excellence in neurological care.

Neuropalliative care is an essential and evolving subspecialty that integrates palliative care principles into managing patients with advanced neurological diseases. Conditions like Parkinson disease, amyotrophic lateral sclerosis (ALS), multiple sclerosis (MS), and dementia lead to symptoms that profoundly affect quality of life for patients and caregivers alike.[1] Neuropalliative care aims to enhance quality of life through holistic, patient-centred approaches, addressing physical, psychological, social, and spiritual needs.[2]

Outlined here are five key practice areas in neuropalliative care: symptom management, communication and goals of care, psychological and caregiver support, ethical decision-making and advance care planning, and a multidisciplinary approach. These components establish an effective neuropalliative care model.

1. Symptom management

Symptom management is foundational to neuropalliative care. Patients with neurodegenerative diseases experience various symptoms, including pain, fatigue, dyspnoea, dysphagia, cognitive decline, mood disorders, and sleep disturbances.[3] Each disease presents its unique spectrum of symptoms, necessitating a tailored approach. For instance, ALS patients may face muscle weakness, respiratory issues, and communication difficulties, while Parkinson disease patients contend with tremors, rigidity, and autonomic symptoms. Clinicians often employ both pharmacological and non-pharmacological approaches, with support from multidisciplinary teams.[1]

Pain management: Neuropathic pain is common, particularly in conditions like multiple sclerosis and advanced Parkinson

disease. Antiepileptics or antidepressants may be effective, while opioids might be necessary for some patients.[4]

Spasticity and mobility: Spasticity in ALS, MS, or post-stroke conditions can be managed with medications such as baclofen or tizanidine, alongside physical therapy to improve mobility.

Cognitive symptoms: Cognitive impairment in conditions such as Alzheimer disease and frontotemporal dementia is managed with cholinesterase inhibitors and memantine, though supportive care plays a significant role.[2]

2. Communication and goals of care discussions

Effective communication is essential in neuropalliative care, especially as progressive conditions such as MS involve prolonged disease courses. Goals of care discussions help align treatment with patients' values, focusing on quality of life rather than extending suffering.[1] These conversations should start early and evolve with disease progression, covering prognosis, interventions (e.g., feeding tubes, tracheostomy), and patient preferences.

These discussions prepare patients and families for transitions from curative to palliative or hospice care, offering guidance and support during challenging times.[4]

3. Psychological and caregiver support

Psychological support is crucial for managing the emotional strain on both patients and caregivers. Neurodegenerative diseases often bring about depression, anxiety, and grief due to functional decline and loss of independence. Counselling, cognitive-behavioural therapy, and pharmacological treatments are important components of neuropalliative care.[2]

Caregivers, often family members, bear significant responsibilities and may experience burnout. Recognizing the caregiver's role, neuropalliative teams provide education, emotional support, and respite care to ensure caregivers can sustain their health and caregiving capacity.[4]

4. Ethical decision-making and advance care planning

Patients with progressive neurological diseases often encounter complex ethical decisions, especially surrounding life-sustaining interventions such as mechanical ventilation and artificial nutrition. Ethical principles, including autonomy, beneficence, non-maleficence, and justice, guide these decisions. Advance care planning (ACP) is essential, particularly for patients who may lose the ability to communicate their wishes as the disease advances.[1]

For example, ALS patients may need to decide early about tracheostomy and long-term ventilation, while those with dementia might consider nutrition and hydration options as dysphagia progresses.[3]

5. The multidisciplinary approach

The complexity of neurological diseases demands a multidisciplinary approach. Neuropalliative care teams generally include neurologists, palliative care specialists, physical and occupational therapists, speech and language pathologists, social workers, psychologists, and spiritual care providers. This collaborative approach addresses physical, emotional, social, and spiritual needs through coordinated expertise.[3]

For instance, physical therapists manage mobility and spasticity, speech pathologists address dysphagia, and spiritual care providers support patients and families with existential concerns.[4]

Neuropalliative care prioritises quality of life by managing symptoms and providing patient-centred care. Focusing on these five key practice points: symptom management, communication, psychological support, ethical decision-making, and a multidisciplinary approach helps healthcare providers support patients and families through the challenges of neurodegenerative diseases.

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Neurology Picture Quiz

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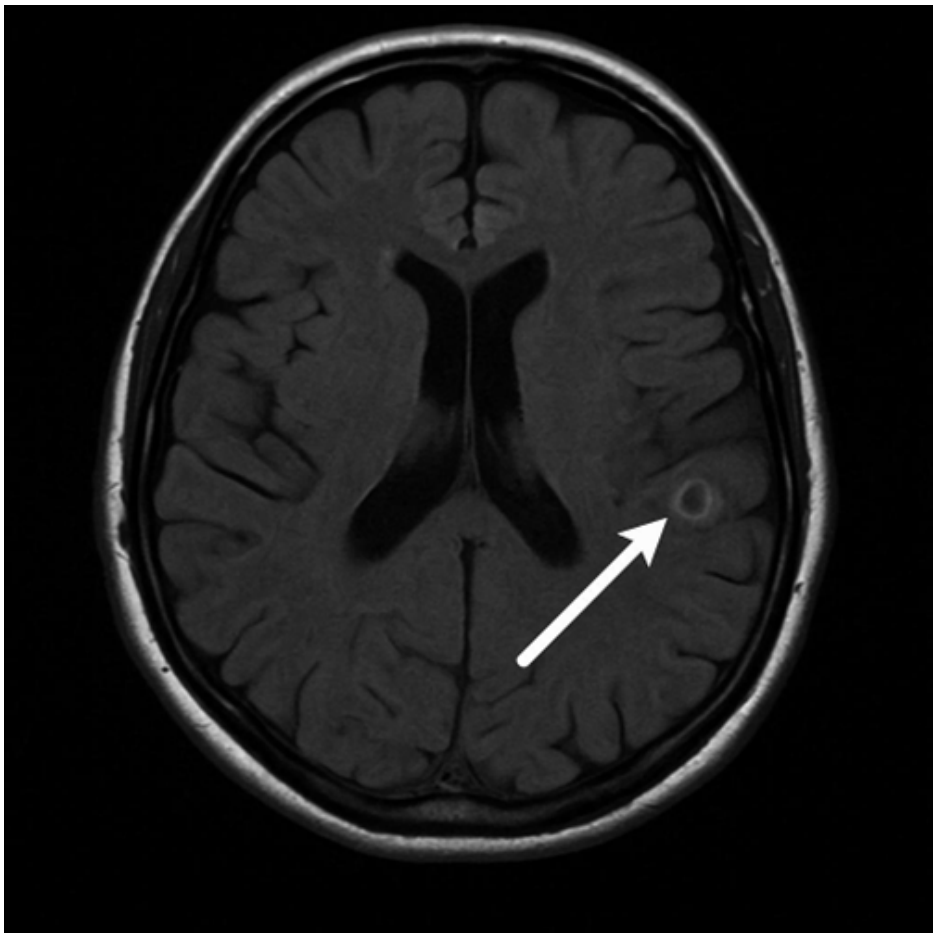
Case 1

This MRI lesion is now considered as a strong predictor and disability worsening of an important chronic immune Neurological disorder and is included in the latest (2024) revision of its diagnostic criteria.

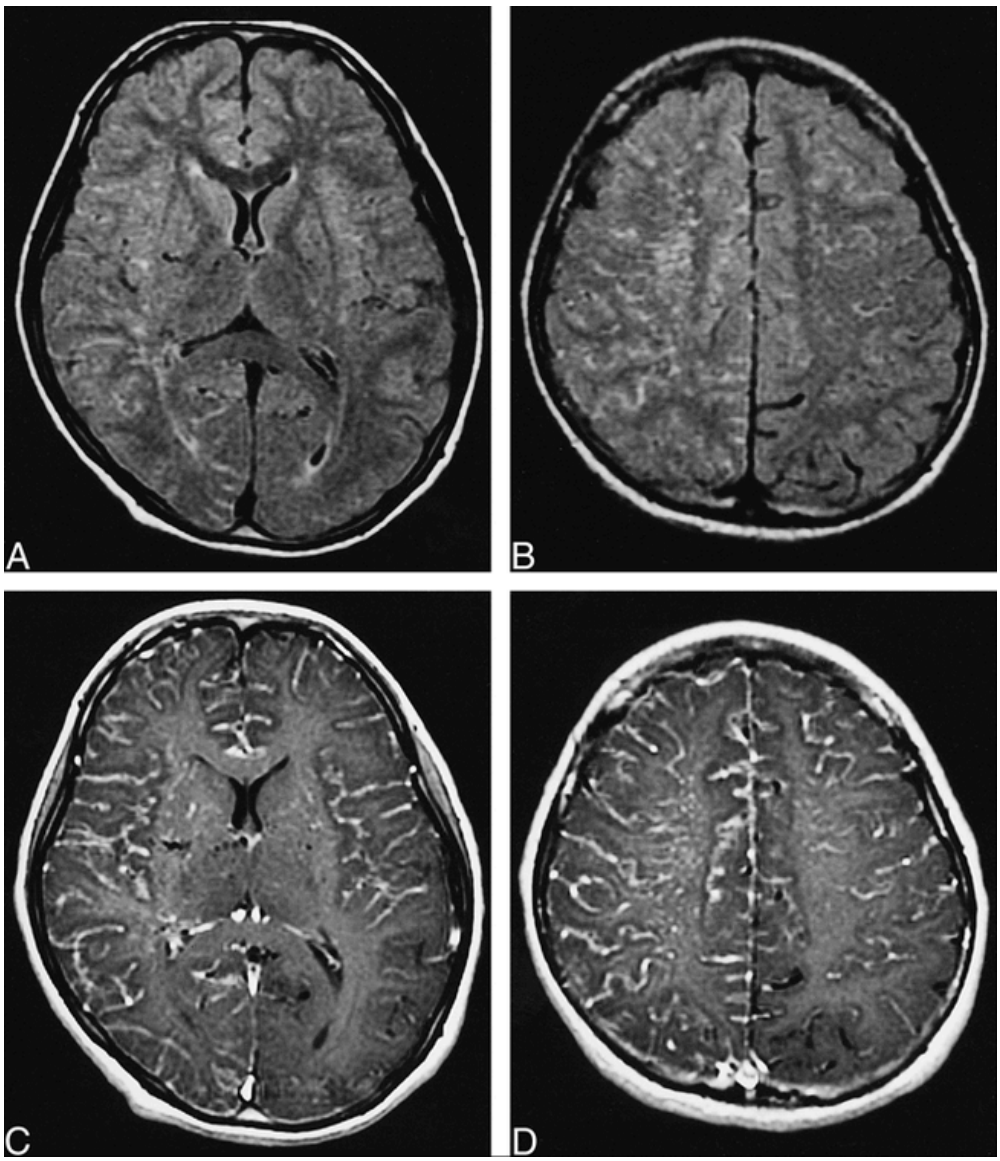
Name a) The lesion b) The Criteria



Case 2

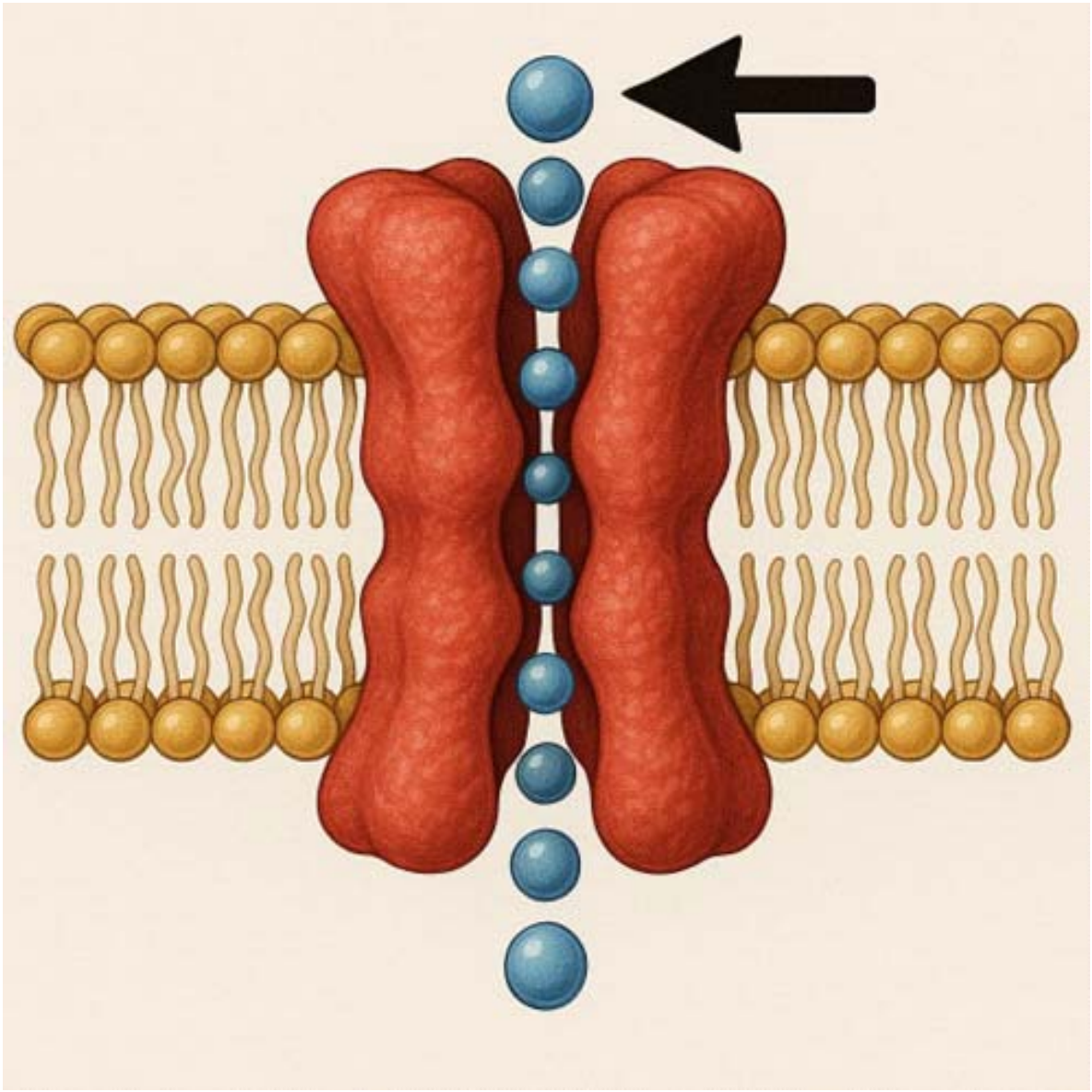
A 15-year-old girl experienced transient motor weakness of the upper and lower left extremities, particularly when she played the trumpet at school. Physical examination revealed no abnormal neurologic findings. FLAIR and contrast T1 images are shown.

a) What is the diagnosis? b) What name is given to the MRI abnormalities seen?



Case 3

This illustrates a channel for water which has many types. Name two types of these channels implicated in important Neurological disorders with their most relevant Neurological disorder.



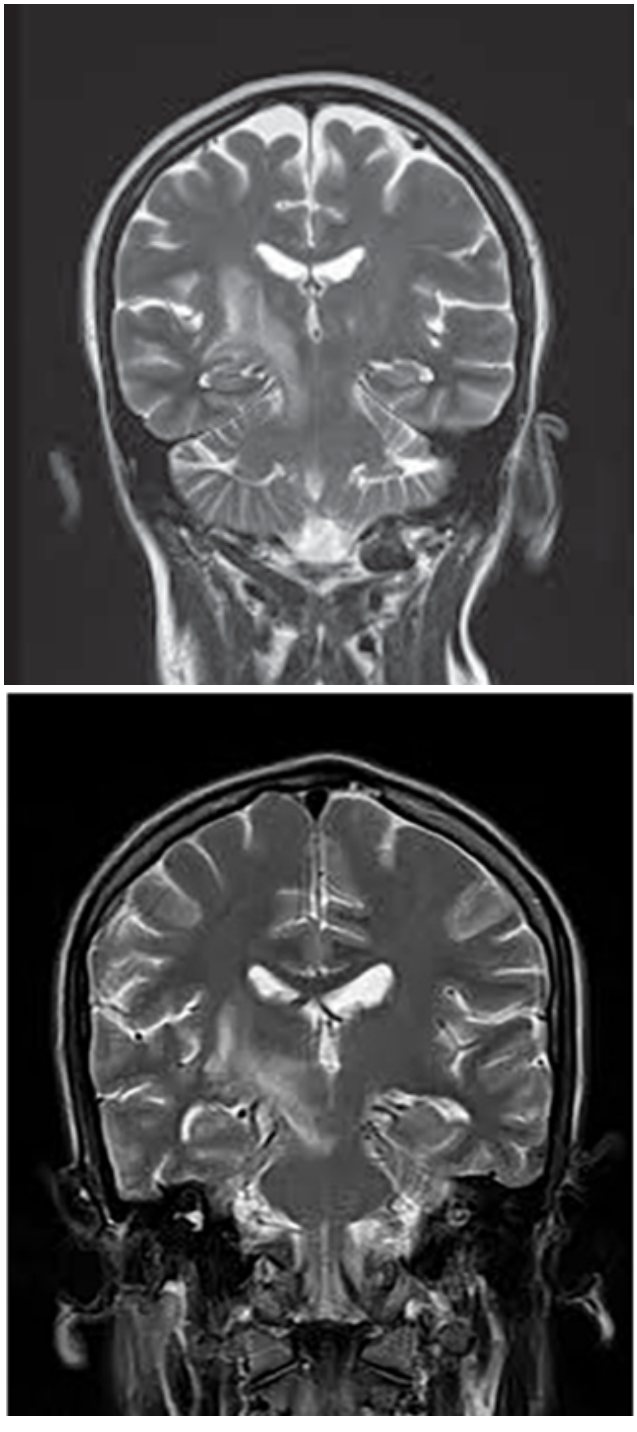
Case 4

A 28-year-old male of African origin presented to the emergency unit with left sided hemiparesis. His family reported a gradual onset of symptoms over an 8-to-10-day period with headaches and left sided numbness affecting his arm and leg. The family reported that he was previously healthy.

On examination 2 ulcers (0.5cm and 1cm) were detected on the scrotum. The cranial nerve examination revealed anisocoria with a dilated and non-reactive right pupil, limited adduction of the right eye and a discrete skew deviation. No uveitis was noted. He had a severe, left sided hemiparesis with facial nerve involvement and a severe left sided limb ataxia. These findings were suggestive of an extensive brain stem lesion or multiple lesions with brainstem involvement.

a) What is the diagnosis?

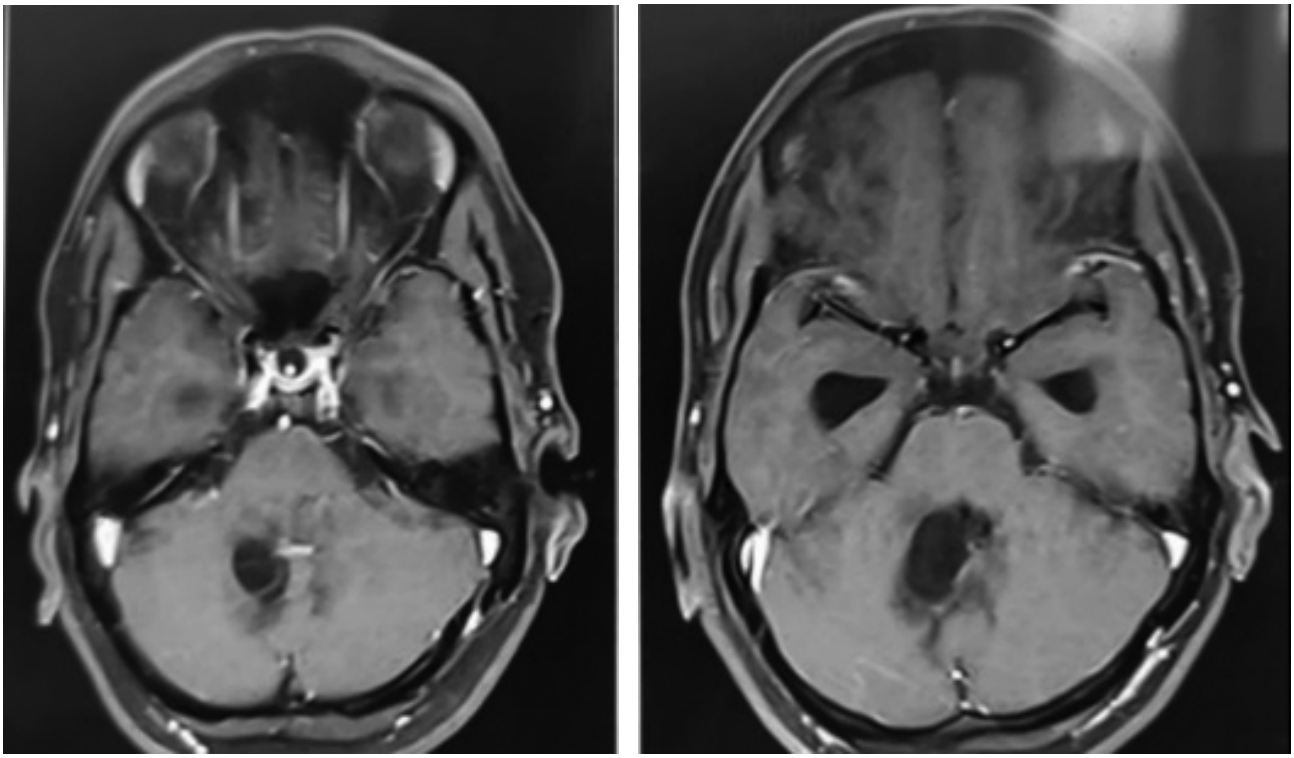
b) What is the name given for the MRI abnormality seen?



Case 5

A 25-year-old woman presented with 2 months of episodic vertigo, vomiting, and headache triggered by abrupt head movements, lasting from a few minutes to 1 hour. She was asymptomatic between the attacks. Fundoscopy showed papilloedema.

a) What is this syndrome? b) What is the likely cause?



Neurology Picture Quiz Answers

Case 1

a) Paramagnetic Rim Lesion (PRL)

b) Mc Donald Criteria

Case 2

a) Moya Moya disease

b) Ivy sign

MRI shows increased signal intensity in bilateral subarachnoid spaces and perivascular spaces on T2-weighted fluid attenuation inversion recovery (FLAIR) MRI.

The ivy sign refers to the MRI brain appearance of patients with moyamoya disease or moyamoya syndrome. Prominent leptomeningeal collaterals result in high signal on FLAIR due to slow flow and vivid contrast enhancement on post-contrast T1. The sign may be more reliably seen on post-contrast T1 than FLAIR. The appearance is reminiscent of the brain having been covered with Ivy.

On level ground ivies remain creeping, not exceeding 5-20 cm height, but on surfaces suitable for climbing, including trees, natural rock outcrops or man-made structures such as quarry rock faces or built masonry and wooden structures, they can climb to at least 30 m above the ground.

Case 3

a) Aquaporin Type 1 – Idiopathic Intracranial hypertension

b) Aquaporin Type 4 – Neuromyelitis Optica

Case 4

a) Neuro-Behcet's disease.

b) Waterfall sign or Cascade sign.

The "waterfall sign" on a brain MRI is a radiological finding characterized by a continuous, high-signal intensity area on T2-weighted or FLAIR sequences that extends from the thalamus and/or internal capsule down to the ipsilateral midbrain. This sign is most commonly associated with neuro-Behcet disease but has also been reported in other conditions such as multiple sclerosis (MS), neuromyelitis optica spectrum disorder (NMOSD), myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD), and glioma.

Case 5

a) Brun's syndrome b) Neurocysticercosis

Brain MRI showed obstructive hydrocephalus and a cystic lesion in the fourth ventricle. Symptoms subsided after cyst excision; a histopathologic diagnosis of neurocysticercosis was made. This clinical picture matches the Bruns syndrome, due to a mobile ventricular mass producing episodic hydrocephalus on changing head posture.

Bruns syndrome is a rare but serious presentation of intraventricular neurocysticercosis, caused by the pork tapeworm larvae (Taenia solium) lodging in the brain's ventricles. It is characterized by sudden, severe headaches, vertigo, and vomiting triggered by abrupt head movements, caused by a cyst temporarily obstructing cerebrospinal fluid flow, leading to a "ball-valve" effect.