

Parietal lobe lesions, in previously normal adults, can cause cognitive deficits of: agnosias – neglect syndromes of illness; body image of self and others; higher order sensations, graphesthesia; motor imagery, self-reflection episodic memory retrieval, praxis, empathy, emotional content of speech. Degree and timing of recovery of each lost function, if it occurs, varies.

An ill person may freely withhold consent, though the converse does not necessarily hold: the giving of consent does not mean a valid choice was made – if awareness was impaired. Capacity, and ‘informed’ consent, especially in parietal lesions, for fully understanding implications of a possible future outcome – which, in the main, is what matters – is very difficult to establish with current neuropsychological tools. This leaves only the specificity of consent possible. Later recovery from the lesion, may allow for valid consent.

Conclusion Medico-legally, these provocative challenges will only increase in the future with an ageing population and more stroke survivors. The questions are not merely hypothetical: difficulties also arise in the spectrum of minimal cognitive impairment to dementia, where the burden of disease may fall on different cerebral lobes.

2422 PARRY ROMBERG SYNDROME/PROGRESSIVE FACIAL ATROPHY (PRS/PFA)-WHO AND HOW OFTEN TO FOLLOW UP

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Objective Justify multi-disciplinary team policy for Follow-up

PRS/PFA is a rare condition with variable presentation though progressive unilateral wasting of the face is the hallmark. It affects females more than males and the left side more than the right and may present to dermatologists, dentists, plastic surgeons, rheumatologists or neurologists. Cause is unclear and symptoms are treated on their merit. Mood is commonly affected and consensus is developing that it should be managed in a multi-disciplinary team setting, including psychologist.

An 18yr old female presented with mild left face wasting. Neurologic examination revealed a mild scoliosis and no morphea.

Childhood PRS/PFA is managed closely with whole-family involvement. Extra-cranial manifestations (ECM) occur in over 40%. Findings of: progressive intra-cranial atrophy, T2 hyperintensities, dementia, epilepsy, calcification, bleeding, trigeminal neuralgia, headaches, movement disorders; numerous eye complications; dental, Temporo-mandibular joint, joint and skin (morphea and sclerosis) – can arise at any time from diagnosis.

As a rare condition, with limited physician exposure, protocols for review differ for each specialty. With around a hundred ECM possible – spread over different fields of expertise, a unified approach would make for efficient use of time and resources over this long-term disease process. Progression of PRS can be expected mainly in the first 10 years. Recurrences and progression, after years of stability, have been reported.

Conclusion Recommendations have been made for: team review each 3–6 months for the first 10 years; MRI each 12–24 months; eye review each 6 months; 3D photography each 6 months

2423 ADMINISTRATION OF SUBCUTANEOUS IMMUNOGLOBULIN (HIZENTRA®) IN THE HOME SETTING THROUGH THE CSL BEHRING CARES PATIENT SUPPORT PROGRAM FOR PATIENTS WITH CIDP

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Objectives Treatment of CIDP includes both intravenous immunoglobulin (IVIg) and subcutaneous immunoglobulin (SCIg). To enable patients with CIDP and who have been prescribed Hizentra® to administer SCIg competently and confidently in the home setting, the CSL Behring CARES™ patient support program, (managed by independent provider, Aesir Health) was made available. An analysis of the first cohort of patients is presented here.

Method Assessment of the program via patient competency in SCIg self-administration.

Results 110 patients with CIDP enrolled in CARES™, ranging from 22 – 90 years old. Average weekly dose of 17.4 g of Hizentra® and overall average dose/kg of 0.22g was administered. Three patients were administering their weekly dose over 2 infusion sessions. Most patients were switched to SCIg following stabilisation with IVIg (2 exceptions commenced on SCIg pre-NBA funding; 1 enrolled from a clinical trial). Initial training session(s) were either via treating hospital or directly from Aesir Health nurses in-home. On average, patients became competent in home self-administration after 1.9 visits (Range, 1–9).

Conclusion A broad age group of patients with CIDP are suitable for home-based therapy with Hizentra®. These patients can be effectively transitioned from hospital-based Ig treatment, to weekly home-based therapy with Hizentra® using the CSL Behring CARES program to help them confidently and competently transition. Further analyses of CARES are planned that will determine how to best continue to support patients with their ongoing home-based treatment.

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2425 REVISITING THE DIAGNOSIS OF CEREBRAL PALSY – WHEN, WHERE AND WHY?

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Objective Argument for complete diagnosis and separating by aetiology.

Cerebral Palsy (CP) has been an umbrella term capturing persons with motor development problems traceable to prenatal and perinatal insults to neurodevelopment. Apt arguments are made for ‘splitting’ and ‘lumping’ diagnostic labels in CP.

A 46 y/o female with Congenital Rubella Syndrome (CRS) presented, post head-strike, with imaging that revealed a prenatal right parietal infarct, and colpocephaly. She exhibited a smaller, spastic, right upper limb, bilateral congenital deafness, different partial colour to the right iris. She had intellectual impairment.

She fulfilled the criteria for monoplegic CP.

As new details of neurodevelopmental processes emerge, the original concept of CP (1843, Little; Osler; Freud), with minor modifications since that time, warrants close reflection.

Arguments by ‘lumpers’ seek to continue current practices for surety of surveillance, rehabilitative, prognostic and financial equipoise purposes: this merits review. Modern-day diagnoses by genetics aid reproductive plans; rehabilitation via virtual therapies relying on vision and hearing; artificial limbs, robotics and the application of nanotechnology for monitoring and mobility purposes – have altered the landscape in which modern CP is contextualized.

Conclusion CP is multi-dimensional: so many trajectories can now be clearly specified, quantified and ameliorated. There are cogent arguments for specifying causes as far as possible (‘splitting’), with specific interventions for each aetiology (including financial), naturally flowing.

2427 RIGHT PARIETAL STROKE: WHAT THEY DON'T KNOW CAN'T HURT THEM!

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Objective Describe a perinatal parietal stroke with emotional anosognosia, presenting in adulthood.

A 46 years old female presented with imaging showing a remote right parietal infarct (superior and inferior lobules), and colpocephaly. There was a history of Congenital Rubella Syndrome (CRS), cognitive impairment and frequent near-miss motor vehicle accidents. She self-reported being an ‘unsettled child’ – in sleep and education. She had difficulty in social situations interpreting others’ intentions toward her, even her husband’s. She had difficulty with child-rearing.

Perinatal strokes occur between 20weeks gestation and 28 days postnatally, presenting catastrophically or with milder Cerebral Palsy-like picture. Asymptomatic cases may go unrecognized for many years: one cause of an ‘asymptomatic’ stroke is a right parietal lesion with ‘anosognosia’.

The parietal lobe is at the cross-roads of vision, hearing, sensation and is involved with mediating self-awareness. Right Parietal strokes can present with difficult to characterize spectrums of misperceptions in vision, hearing and emotion. However, two further considerations apply here: 1) the disturbance of parietal function had occurred preceding the development of Parieto-cortical (short and long) fibre connections, and, 2) long fascicular tracts traversing through that parietal lobe have had their connections interrupted, secondarily affecting inter-hemispheric functions.

Conclusion A prenatal right parietal stroke is rare enough that neuropsychological test ‘normal values’ would be difficult to interpret. Patients who present with longstanding neurological issues should be imaged at least once as an adult. Validated neuropsychological testing for the right parietal lobe needs to be developed to better understand this debilitating condition.

2428 ‘AN UNNERVING PROBLEM’, A CASE OF SEVERE RAPIDLY PROGRESSING POLYNEUROPATHY IN AN ELDERLY LADY

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Background

- The patient is A 88-year-old lady who initially presented for workup of a pelvic mass but subsequently found to have a rapidly progressing weakness of both legs and hands.
- 5 weeks ago, the patient was independent with all activities of daily living. Currently, the patient had bilateral foot drop worst on the left, bilateral wrist drops, impaired pain and proprioception with preservation of reflexes on clinical examination.

Investigation and Treatment

- On initial presentation, patient had raised ESR 110 and raised CRP 115 with unclear cause. Septic screen and cultures were negative. Autoimmune screen was negative.
- CSF studies including cultures, chemistry, oligoclonal bands and cytology were normal
- Neuroimaging including MRI scan of the entire spine and CT brain were normal
- Significantly raised paraproteins but polyclonal and non-specific.
- Nerve conduction studies confirmed severe sensory axonal polyneuropathy, but interpretation greatly limited by patients’ ability to tolerate the examination. Coexisting motor neurone involvement could not be excluded on current study.
- Was treated with IVIG using CIDP protocol with some improvement in patient’s motor function.

Goals and learning points of presentation.

- To highlight the challenges and difficulty in managing a patient with rapidly progressive polyneuropathy in the geriatric age group with unclear cause
- To initiate an open discussion regarding the approach to diagnosis and management of these patients

2433 COVID VACCINATION-RELATED EXACERBATION OF SEIZURES IN PATIENTS WITH EPILEPSY

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Objectives 12 months since the implementation of the COVID vaccination, over 94% of the Australian population over 16 years old are fully vaccinated. Although vaccines are generally safe in persons with epilepsy (PWE), seizure-like events are a known complication of vaccinations, including COVID vaccines. This study assessed the rate of COVID vaccination-related exacerbation of seizures in PWE.

Methods Adult PWE who had received at least one COVID vaccine were prospectively recruited at the epilepsy clinic between June 2021 and February 2022. Patient demographics, including epilepsy history, vaccination details and side effects were recorded. The rate of seizure exacerbation, defined as within one week of vaccination, was assessed.

Results 364 PWE received the COVID vaccine, with 352 patients (97%) receiving two doses, with 73% receiving the Pfizer vaccine as their initial dose. 31% of patients were 12-months seizure free at baseline. The median number of anti-seizure medications (ASM) was 2, with 65% of patients on 2 or more ASM. Most patients (62%) had focal epilepsy. 10