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Review

# Epilepsy in paediatric patients with Parry-Romberg syndrome: A review of the literature

Ruben Rocha[a,*], Marios Kaliakatsos[b]

[a] Centro Materno infantil do Norte, Centro Hospitalar Universitário do Porto, Porto, Portugal
[b] Paediatric Neurology, Great Ormond Street Hospital for Children, London, United Kingdom

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ABSTRACT

*Background:* Parry–Romberg syndrome (PRS) is a rare disorder characterized by unilateral slow progressive facial atrophy that can be associated with neurologic manifestations, namely seizures. There is scarce data about seizures in paediatric patients with PRS. The aim of our work was to clarify the clinical features of paediatric patients with PRS and seizures.
*Methods:* We performed a literature review based on a literature search using PubMed and EMBASE databases. We included original articles in which the main diagnosis was PRS and the patients were 17 years old or less when the first seizure occurred.
*Results:* We included 40 patients. Most of the patients had previously normal development and had their first seizure in the first decade of life. Neurologic examination was abnormal in 56 % of patients. Seizures are typically focal, frequently with impaired awareness, and became refractory in about 40 % of patients. Few patients have generalized seizures. On electroencephalogram, epileptic discharges are generally focal, on the same side as the facial atrophy, without a predominant cerebral lobe localization. Brain MRI is almost always abnormal, typically with T2 subcortical hyperintensities, and sometimes brain atrophy or calcifications. In addition to the classic antiepileptic drugs, immunosuppressive drugs should be considered as potential epilepsy treatment.
*Conclusion:* To the best of our knowledge, this is the first review dedicated to the characteristics of paediatric patients with PRS and epilepsy. Seizures are usually focal, became refractory in 40 %, and have a significant impact on the quality of life and neurodevelopment of patients.

## 1. Introduction

Parry–Romberg syndrome (PRS) was first described by Parry in 1825 and later by Romberg in 1846 [1]. PRS is a rare disorder characterized by unilateral slow progressive facial atrophy that can affect all tissues, from the skin to the osteocartilaginous structures. It is sometimes

associated with a peculiar skin lesion called “en coup de sabre”– a subtype of localised scleroderma [2]. Overlap exits between these two entities, and in the literature there is still discussion as to whether they represent variants of the same disease with different manifestations (involving the skin in scleroderma “en coup de sabre” and subcutaneous tissue, muscle and bone in PRS).

The real incidence of PRS has not been ascertained so far and the aetiology is uncertain [3]. Autoimmunity, trauma, infection, vascular and autonomic dysregulation have all been suggested, but autoimmunity is the leading theory [3].

The first symptoms frequently start in the first two decades of life with progressive hemifacial atrophy of craniofacial tissues inferior to the forehead, typically involving dermatomes of one or more branches of the fifth cranial nerve [3].

PRS has been associated with various systemic manifestations, principally neurologic, ophthalmologic and maxillofacial [4]. Neurological manifestations can include seizures, headaches, movement disorders or neuropsychological symptoms. In a worldwide survey, about 11 % of PRS patients had coexistent epilepsy [5]. Knowledge about the seizure types and epilepsy of paediatric patients with PRS is limited. The aim of our work was to clarify the clinical features of paediatric patients with PRS and seizures.

## 2. Methods

We performed a literature review based on a literature search using

* Corresponding author at: Centro Materno Infantil do Norte, Centro Hospitalar Universitário do Porto, Largo da Maternidade de Júlio Dinis, 4050-651 Porto, Portugal.
*E-mail address:* rubenrocha@gmail.com (R. Rocha).

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**Table 1**
Summary of the characteristics of patients included in the review.

| Author | Age | Age of onset | Sex | Affected cutaneous side / frontal skin lesion | Seizures type | EEG discharges location | Neuro exam | Brain imaging (MRI) | Treatment | Seizures outcome |
|---|---|---|---|---|---|---|---|---|---|---|
| Sotgiu [10] | 5 | 2 | M | Left / yes | Nonconvulsive status epilepticus Dyscognitive seizures | Spike and slow-wave complexes on left temporal region. | Normal | FLAIR hyperintensities in left mediotemporal areas | Clobazam Methylprednisolone, methotrexate | Seizure free Hyperactivity |
| Zakkiriak [11] | 14 | 14 | M | Left / yes | Right focal- aware | Epileptiform abnormalities in left fronto central area | Normal | Increased signal intensities in the left hemisphere, mainly in the frontal lobe, with areas of microhemorrhages and calcifications | Oxcarbazepine, prednisolone, methotrexate | Well controlled |
| Zakkiriak [11] | 7 | 5 | F | Left / Yes | Right focal - bilateralization | Spike activity over the left centro temporal area | Normal | Hemiatrophy of the left cerebral hemisphere with hyperintensity (FLAIR) in the left temporal lobe | VPA, oxcarbazepine, LEV, prednisolone, methotrexate | Well controlled |
| Anderson [12] | 10 | 8 | F | Left / yes | Focal? | Not described (temporal lobe epilepsy) | | Left frontal and parietal intraparenchymal microbleeds | LEV, steroids and methotrexate (the later improved seizures) | 2-year period with only one seizure |
| English [13] | 12 | 11 | M | Left / yes | Right focal - awareness- bilateralization | Epileptiform discharges in left cerebral hemisphere Moderately increased amplitude of the background activity | Sensory examination revealed decreased light touch over the right side | Patchy left supratentorial white matter T2 hyperintensity, as well as cystic changes in the left basal ganglia, microhemorrhages in white matter. Contrast-enhanced images: diffuse superficial and deep perivascular nodular enhancement in a distribution suggestive of diffuse venous inflammation. Arterial spin labeling perfusion: globally diminished flow within the left cerebral hemisphere. Gyriform calcifications | LEV, methotrexate and prednisolone | Recurrent seizures at the time of last evaluation |
| Yanagishita [14] | 5 | 4 | F | Right / yes | ? | ? | | Cortical atrophy and calcifications in the right frontal lobe. T2 hyperintense areas in the white matter of the right frontal lobe. | | |
| Somer [15] | 11 | 5 | F | Left/yes | Focal - impaired awareness | | Pupil asymmetry | T2 hyperintense signals in left hemisphere | Methotrexate, multiple anti-epileptic drugs | Refractory Epilepsy |
| Doolittle [16] | 28 | 14 | F | Right / yes | Generalized | Normal | Normal | Nodular gray matter heterotopia adjacent to the frontal horns of both lateral ventricles | | |
| Doolittle [16] | 16 | 6 | M | Left / no | Generalized | Right posterior focal slowing and epileptiform discharges lateralized to the right hemisphere | Nystagmus, visual impairment, Wide-based gait and trouble with tandem gait | Contralateral deep and subcortical white matter T2 hyperintensity Contralateral parietal cortical T2 hyperintensity Diffuse contralateral cerebral atrophy Ipsilateral parietal atrophy | | |
| Garófalo Gómez [17] | 7 | 2 | M | Left/ yes | Focal- impaired awareness | | | CT scan - calcification in the left brain hemisphere and a hypodense area in the head of the left caudate nucleus | CBZ, clobazam | Seizure controlled |

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**Table 1** (*continued*)

| Author | Age | Age of onset | Sex | Affected cutaneous side / frontal skin lesion | Seizures type | EEG discharges location | Neuro exam | Brain imaging (MRI) | Treatment | Seizures outcome |
|---|---|---|---|---|---|---|---|---|---|---|
| Dupont [18] | 24 | 4 | F | Right / yes | Focal - impaired awareness | ? | Normal | Gyration and white gray matter demarcation were abnormal in the anterior right frontal lobe and temporal lobe. In same area there were white matter T2 hyperintensities and heterogeneous cyst-like zones in the right basal ganglia area | | Refractory epilepsy |
| Madruga [19] | 11 | 3 | F | Left / no | Focal? | Epileptiform discharges in right cerebral hemisphere | Normal | Normal | CBZ | Seizures controlled with medication |
| Kakisaka [20] | 26 | 16 | M | Left/yes | Generalized Left Focal clonic EPC | | Left hemiparesis. Babinski sign at left | Right hemispheric atrophy, most prominent in peri-rolandic and insular regions | VPA, zonizamide, immunoglobulin | Refractory epilepsy |
| Longo [21] | 6 | 6 | M | Left / no | Left focal - aware | Repetitive slow waves over the left frontotemporal and parietal areas, sometimes intermingled with spikes and waves more evident during sleep | Normal | Mild extended T1 and T2 signal changes in the white matter of the left frontal region. Increased atrophy of the left hemisphere, particularly the frontotemporal region | Several antiepileptic drugs Steroids Immunoglobulin and phenobarbital reduced seizures frequency | Refractory epilepsy, Learning difficulties |
| Seifert [22] | 23 | 14 | M | Left, no | Focal - impaired awareness | | | Left frontal lesion (hyperintensity FLAIR) | Many antiepileptic drugs Surgery (type?) prednisolone Immunoglobulin (best) | Refractory epilepsy |
| KaciñsKi [23] | 11 | 10 | M | Left, no | Focal – impaired awareness | Sharp waves and single sharp and slow wave complexes in right posterior region | Intermittent right VI nerve palsy | Normal | CBZ, oxcarbazepine | Good control of seizures |
| Moseley [9] | 23 | 5? | M | Left / yes | Left focal - aware | Discharges in right parietal occipital region | | Right cerebral/cerebellar and left occipital atrophy. T2 hyperintensities within the right cerebral white matter | Several antiepileptic drugs Rituximab improved seizures | Epilepsy improved after rituximab |
| Qureshi [24] | 10 | 10 | M | Left / no | Right focal - impaired awareness | Epileptiform activity in centrotemporal areas | Right hemiparesis | Atrophy of the left hemisphere, hypoplastic left internal carotid artery. Left cerebral arteries were hypoplastic. | | |
| Satori [25] | 9 | 6 | M | Left / yes | Left focal – impaired awareness Status epilepticus | | Normal | Cortical and subcortical areas of hyperintense signal on T2 weighted and FLAIR sequences in the paramedian and basal regions of left frontal lobe and left temporal lobe | Clobazam, oxcarbazepine Prednisolone and methotrexate | Good seizure control. Learning disabilities improved with prednisolone and methotrexate. |
| Verhelst [26] | 8 | 2 | F | Right / yes | Focal - aware | Right temporal focus with sharp slow-waves | Normal | Hippocampal atrophy on the right | Several antiepileptic drugs VPA, nitrazepam and topiramate (best combination -seizure free) | Seizure free with 3 antiepileptics, after a period several seizures, cognitive decline |
| Carreño [27] | 9 | 3 | F | Left / no | Right focal. EPC. | Continuous slowing and multiregional sharp waves over the left hemisphere. One hundred seizures recorded arising from the left frontal region | Hemiparesis | Atrophy of the left hemisphere | Surgery (Functional hemispherectomy) | Seizures decreased dramatically after surgery |

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**Table 1** (*continued*)

| Author | Age | Age of onset | Sex | Affected cutaneous side / frontal skin lesion | Seizures type | EEG discharges location | Neuro exam | Brain imaging (MRI) | Treatment | Seizures outcome |
|---|---|---|---|---|---|---|---|---|---|---|
| Haldar [28] | 17 | 16 | F | Right / yes | Left focal and generalized | Normal | Left hemiparesis | Focal atrophy in right frontoparietal cortex | Prednisolone improved seizures | Seizures stopped with steroids, only 1 seizure thereafter |
| Sommer [29] | 14 | 3 | F | Right/ no | Focal | Overlying beta activity | Normal | Frontoparietal subcortical hyperintense lesion in FLAIR and T1 | | |
| Sommer [29] | 14 | 7 | F | Left / yes | Generalized tonic-clonic | Normal | Normal | Normal | | |
| Sommer [29] | 26 | 6 | F | Bilateral/ yes | Generalized tonic-clonic | Frontotemporal theta-delta activity | Palsy of cranial nerves 3,4,6,7 | Frontoparietal subcortical hyperintense lesion in FLAIR and T1 | | |
| Sommer [29] | 26 | 4 | M | Left/ yes | Generalized tonic-clonic | Normal | Fixed pupil | Ventricular dilatation, subcortical calcification | | |
| Paprocka [30] | 10 | 6 | F | Left / yes | Right Focal | General paroxysmal changes with variable hemisphere predominance. | Right hemiparesis | Hyperintense lesion in left pericentral lobulus | Surgery (Cortical resection and multiple subpial resections) | Refractory epilepsy even after surgery, Cognitive decline |
| Sathornsumetee [31] | 4 | 2 | M | Right / yes | Left focal | Right temporal slowing and occasional spikes | Right facial palsy, right oculomotor palsy | Minimal cerebral atrophy on the right, asymmetry of hippocampal structures, and a right frontal lobe lesion | Methotrexate and prednislone, later immunoglobulin and cyclophosphamide | Refractory epilepsy, Died |
| Chbicheb [32] | 17 | 5 | M | Right / yes | Left focal – impaired awareness | Slow right occipital activity | | Right occipital atrophy and dilation of the occipital horn of lateral ventricle. Right occipital calcifications. | Several antiepileptic drugs Surgery (1st - right occipital lobectomy and partial right parietal resection; 2nd right parietal resection and anterior disconnection) | Refractory epilepsy, cognitive decline |
| Chbicheb [32] | 5 | 5 | F | Right / yes | Focal- impaired awareness | Spike-wave and theta activity in right temporoparietal region | | Moderate ventricular dilation | Phenobarbital Vigabatrin Topiramate Steroids | Refractory epilepsy, Learning difficulties |
| Sandhu [33] | 4 | 3 | M | Left / no | Focal - impaired awareness | ? | Normal | Subdural hygroma | phenytoin | |
| Shah [34] | 7 | 7 | M | Right / yes | Left focal. EPC Generalized. | Intermittent, low to medium voltage, polymorphic slow wave activity of the right hemisphere. Rare sharp wave activity emanating from the right frontal region | Left hemiparesis and deep tendon reflexes brisker on the left. fine motor movements moderately impaired in the left hand. | Atrophy of right frontal and parietal lobe. T2 and FLAIR in the right frontal cortex with minimal white matter involvement | VPA, CBZ, clonazepam, oxcarbazepine Surgery (Right Hemispherectomy) | Seizure free after surgery |
| Castañeda- Reyna [35] | 20 | 10 | M | Left / no | Focal – impaired awareness | | Normal | Extensive cystic change in deep white matter of left temporo-occipital region | CBZ, VPA, gabapentin | Died in status epilepticus |
| Yano [36] | 3 | 3 | M | Left/ yes | Right focal - impaired awareness | Slow-wave bursts lasting 1–2 s predominantly over the left frontocentral area during both waking state and sleep | | Focal areas of T1 and T2 prolongation in the frontal white matter adjacent to the left cingulate semiovale. Discrete linear calcifications | VPA, CBZ | Seizures controlled |
| Goldberg-stern [37] | 7 | | F | Left / no | Focal – impaired awareness | Mild left slowing with no epileptiform discharges | Brisk deep tendon reflexes on the left. Disk edema and anterior uveitis | Left frontal T2 hyperintensities, left frontal region. Linear contrast-enhancing lesion in the left frontal lobe | CBZ prednisone and methotrexate | No seizures on CBZ |
| Taylor [38] | 14 | 2 | F | Right / yes | Focal -impaired awareness | Few bursts of high amplitude delta waves predominately on the left | | Calcifications of temporal lobe. | Lamotrigine carbamazepine | Seizures controlled |

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**Table 1** (*continued*)

| Author | Age | Age of onset | Sex | Affected cutaneous side / frontal skin lesion | Seizures type | EEG discharges location | Neuro exam | Brain imaging (MRI) | Treatment | Seizures outcome |
|---|---|---|---|---|---|---|---|---|---|---|
| Takahashi [39] | 30 | 15 | M | Right / no | Focal | Spike discharges predominantly in the right occipital lobe | Left upper temporal homonymous hemianopsia | Multiple cerebral calcifications | | |
| Derex [40] | 15 | 9 | F | Left /no | Focal - impaired awareness | Spike and wave complexes in left hemisphere | Hemiparesis | Porencephaly on left hemisphere, left parietal and frontal calcifications | Vigabatrin, CBZ | Seizures controlled. mild cognitive impairment |
| Miller [41] | 21 | 9 | F | Right / yes | Focal | ? | Partial III nerve palsy | Normal | Phenobarbital | Only 2 seizure not medicated |
| Klene [42] | < 18 | < 18 | F | | Focal | | | Hemiatrophy of brain | | |

VPA- valproic acid; LEV -levetiracetam; CBZ- carbamazepine; EPC – epilepsia partialis continua;? – not specified in original article.

PubMed and EMBASE databases. The main goal of this review was to summarize the clinical findings, diagnosis, clinical course and proposed treatments of paediatric cases of Parry-Romberg and epilepsy.

The search terms were “Parry-Romberg syndrome”, “progressive hemifacial atrophy”, “progressive facial atrophy”, “epilepsy” and “seizure”. An example of a MEDLINE

research query was: (Parry-Romberg syndrome OR progressive hemifacial atrophy OR progressive facial atrophy AND (epilepsy OR seizure)).

We included original articles of retrospective cohort studies, case series and case reports in the English language literature or available translations of international literature. The search has been limited to articles published in last 30 years (from 1st of January 1989 to 1st of December 2019). A cross-check of the references from the original studies was performed in order to identify potential additional papers. The eligibility criteria for inclusion required that the original article refer to PRS as the main diagnosis of the patient, and that the patient was 17 years old or younger when first seizure occurred. Additional neurologic symptoms or diseases (e.g. Rasmussen encephalitis) were not criteria for exclusion. We excluded cases of patients in which the main diagnosis defined by the authors were linear scleroderma or Dyke-Davidoff-Masson Syndrome. Cases of PRS without seizures were excluded. Original articles with poor information regarding epilepsy in PRS were also excluded.

After completion of the searches and excluding the duplicate studies, the initial screening of publications included reviewing titles and abstracts. For documents fitting the inclusion criteria, we collected the following information: first author, publication year, age of PRS diagnosis, age of onset of seizures, seizures types and seizures descriptions, electroencephalogram reports, imaging data and outcome. Descriptive statistic was used to summarise the study results.

## 3. Results

We identified 167 articles with the key search terms and 34 of them met the inclusion criteria. Articles not selected were excluded because important information was missing, the main diagnosis was other than PRS (e.g. linear scleroderma), the patient had a clinical presentation that strongly overlapped with other diseases and age of onset of seizures was greater than 17 years of age.

Most of the papers were case reports and cases series. The maximum number of patients in an article were four. We included 40 patients. Twenty patients (50 %) were female and 20 patients were male. Table 1 summarizes the characteristics of patients included.

Most patients had previously normal development and had their first seizure in the first decade of life. Neurologic examination was abnormal in 56 % (17/30) of patients. Neurologic abnormalities were different in different patients, but the most common reported abnormality was hemiparesis.

Seizures were typically focal in the 85 % (34 /40) of patients and commonly associated with impaired awareness. Some patients experienced evolution to bilateral convulsive seizures and there were 8 patients (22 %) who also or exclusively had primary generalised seizures. The duration of seizure was missing in most articles and no conclusion could be made with regards to this. The seizures tended to increase in frequency with time and became refractory in a significant number of patients. There was data about seizure outcome in 29 patients, and in 11 (38 %) seizures were refractory. Some patients developed epilepsia partialis continua. Table 2 show some descriptions of focal seizures in PRS patients. Electroencephalographic findings included, at least in the early stages of the disease, a normal background rhythm. Epileptic discharges were generally focal, on the same side as the facial atrophy. Epileptiform discharges did not preferentially affect a specific cerebral lobe, and in a significant number of patients discharges involved all of the hemisphere.

Brain imaging (MRI) was abnormal in 36 patients (90 %), only 4 patients had a normal brain MRI. Common abnormalities were T2 hyperintensities, which could be diffuse or localized, and when localized

**Table 2**
Seizure semiology of some patients.

| Author | Seizure semiology |
|---|---|
| English [13] | "a crawling sensation over his right foot that rose through his body and subsequent generalized tonic-clonic seizure"<br>"episodes of right-sided numbness and weakness without secondary generalization" |
| Dupont [18] | "rich set of subjective symptoms, consisting of various vegetative and psychic sensations such such as thoracic pressure, a feeling of warmth, micropsia or macropsia, and gustative or olfactory hallucinations, sometimes followed by complex partial seizures with pale- ness and swallowing." |
| Moseley [9] | "slowness to respond, eye fluttering with speech arrest and rare left leg twitching" |
| Verhelst [26] | "a sensation of pain in the left corner of the mouth and the left arm, rapidly followed on most occasions by twitching of the left corner of the mouth and the left arm." |
| Derex [40] | "rupture of the contact, chewing and complex automatisms, lasting one minute." |

were more frequent in the frontal region. Hemispheric atrophy and calcifications were also were reported as frequent related features.

Little information regarding prognosis was available, but, as already mentioned, seizures became refractory over time in 38 % of patients. These patients had tried various antiepileptic drugs with limited success.

Immunosuppressive drugs used in the treatment of dermatologic condition may also have some impact in seizures control. In at least four patients there was a temporal relation between seizure control and initiation of immunosuppressive drugs.

Surgery in PRS for refractory epilepsy was an option for five patients. Two patients had a significant benefit with regards to seizures. Hemispherectomy was the only surgery type associated with seizure improvement. Less extensive procedures were not associated with a better outcome.

Cognitive decline and learning disabilities were reported and associated with refractory epilepsy. Two patients died, one in status epilepticus.

## 4. Discussion

Debate continues as to whether PRS and linear scleroderma / morphea en coup de sabre are different processes or are part of a single disease spectrum. Currently, there is a tendency to consider them variants of localized scleroderma [6]. In linear scleroderma /morphea en coup de sabre there is normally a cutaneous lesion and skin induration, predominantly affecting the forehead, and sometimes associated with hyperpigmentation and alopecia. Histopathologic features include dermal sclerosis. In PRS there is paramedian atrophy of the face, and the atrophy may extend to the entire hemiface with no overlying skin induration. Histopathological features include atrophy of subcutaneous, fat, muscle and osteocartilaginous tissues [6]. These differences are difficult to separate clinically and overlap occurs in an important percentage of cases. While at present there is no definite answer to this question, we preferred to separate these two entities, whenever it was possible, in order to select a homogeneous sample (and avoid mixing different variants or diseases). For this reason, and because they probably represent a more severe phenotype, we decided to include only patients with the diagnosis of PRS.

Neurologic involvement is frequent among PRS patients. Of the various neurological manifestations seizures are one of most frequent reported in literature [5].

Despite this fact, little has been written in the medical literature about the subject, especially concerning paediatric patients. To the best of our knowledge, this is the first review dedicated to discussing the characteristics of paediatric patients with PRS with epilepsy.

Seizures in PRS can be explained by intracerebral lesions that usually accompany facial involvement of the disease, even in cases where that lesion is not identifiable on MRI. In our review brain MRI frequently revealed abnormalities, often involving the hemisphere ipsilateral to the skin lesions. These lesions appeared as T2 hyperintensities, mostly in the subcortical white matter, but also as brain atrophy or calcifications. These findings were in accordance with the extensive review of Chiu et al. concerning brain imaging in PRS [7].

In a review of neurologic manifestations of PRS, which included adult patients, epilepsy was reported to occur in 60.5 % of patients [8]. Seizures were focal in 50 % of patients and generalized in 43.6 % of the patients [8]. It was estimated that in about a third of patients these seizures are refractory to medication. In our review more than 85 % of paediatric patients had focal seizures. Usually the affected cutaneous site is contralateral to seizure semiology, and ipsilateral with neuroimaging and electroencephalogram findings. Some patients presented with a clinical picture of cerebral unihemispheric atrophy and intractable focal seizures suggestive of Rasmussen encephalitis [9].

Electroencephalogram is reported to be abnormal in most patients with seizures and discharges were mainly localized in frontal or temporal regions [8]. In this review we didn't see preferential involvement of particular cerebral lobes. Some patients have discharges mainly in posterior regions. It is reported that about 10 % of the patients have an abnormal EEG without a past or present history of seizures [8], but in this review we only included patients with seizures.

Immunosuppressive drugs used in the treatment of associated dermatologic conditions may have some impact on seizure control in some patients, and should probably be considered as an alternative to classic antiepileptic drugs.

Although only 5 surgical cases were reported here, patients who underwent hemispherectomy had better seizure control than patients with less extensive procedures. This finding agrees with the concept that lesions visible on brain MRI may be only part of the total area of abnormality.

Given the publication bias and retrospective nature of reports, we are cautious about the conclusions of this review. Deriving epidemiological data from retrospective cases series and reports is difficult and has some associated flaws. There may be a tendency to preferentially publish 'interesting' cases, especially single cases, and this may not be representative of the majority of patients. Most of the articles were not dedicated to neurological symptoms or epilepsy, so information regarding seizure characterization is often limited. However, the fact that most reports were not dedicated to epilepsy may represent a potential strength. Most cases concerned PRS in general (rather than epilepsy in PRS) and as a result the data concerning epilepsy is potentially less likely to suffer from publication bias.

Despite the limitations, this review allowed us to better characterise epilepsy in paediatric patients with PRS, and to gather some preliminary data about outcome. Prospective data collection is urgently needed.

Seizures represent an important component of the burden of the disease and significantly affect both quality of life and neurodevelopment. Neurologic sequelae included motor deficits (such as hemiparesis) and cognitive deficits (ranging from mild learning disabilities to important intellectual disability).

## 5. Conclusion

Paediatric patients with PRS usually develop seizures in the first decade of life. Seizures are predominantly focal with impaired awareness, and tend to became refractory in about 40 % of cases. Seizures seem to affect the quality of life and neurodevelopment of these children. Treatment of seizures include classic antiepileptic drugs, but immunosuppressive drugs and surgery should also be considered.

## Declaration of Competing Interest

None.

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