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Research Article

Paediatric Intracranial Arachnoid Cysts: Single Institutional Review and Outcome

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ABSTRACT

Background: Intracranial arachnoid cysts in the paediatric neurosurgical patient are a common diagnostic quagmire. Though commonly an incidental finding, they are a continuous source of anxiety for both patient's families and the managing clinician. The symptoms often attributed to their presence are also often varied and do range from nonspecific headache or behavioural changes to life threatening hydrocephalus or intracystic haemorrhage. **Materials and Methods:** We present a 6-year retrospective review and analysis of cases of intracranial arachnoid cysts diagnosed at a single centre tertiary care paediatric neurosciences unit. An analysis of medical records and review of images was performed. **Results:** A total of 167 cases were identified over a period of 6 years, with 69% being males and 31% females. Age at diagnosis varied from 1 day of life to 17 yrs. Follow up duration has been up to 11 years. Presentation varied from incidental finding, headaches (7.8%), seizures (2.4%), vomiting (2.4%), behavioural anomalies to decreased level of consciousness (1.2%). On radiological imaging, majority were located in the skull base: middle cranial fossa (55.1%) and posterior cranial fossa (38.9%). Medical and expectant management of these patients was the primary mode of care, with surgical intervention only needed in cases characterized by either hydrocephalus, significant mass effect with features of raised ICP or in cases of intracystic haemorrhage. **Conclusion:** Arachnoid cysts generally have a benign course and can be managed expectantly. In our series, none of the children managed conservatively later on required any neurosurgical intervention. Surgery however has a role in cases presenting acutely with hydrocephalus, significant mass effect or due to bleeding into the arachnoid cyst. Further research and analysis is necessary to elucidate the relationship between arachnoid cysts and complex behavioural / neuropsychiatric conditions in children.

Keywords: Arachnoid cyst, Intracranial cyst, Congenital intracranial cyst, Benign cyst, Brain imaging.

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INTRODUCTION

Arachnoid cysts are localised enlargements of the subarachnoid spaces. These are however isolated from the natural Cerebrospinal Fluid (CSF) flow pathway with the basal cisterns or ventricular system.[1,2] They are also commonly associated with blood vessels running through them. As a result, there is always concern about possible progression in size or intracystic haemorrhage with deleterious consequences. It has been theorised that they form as a result of splitting of the arachnoid layer with a "ball-valve mechanism" responsible for the splitting or the layers and consequent increase in size.[1-3]

With increased availability of neuro imaging, more cases of incidental arachnoid cysts are being reported. The trigger for neuroimaging is also varied may include occasional headaches, migraines, trauma, epilepsy, behavioural issues or screening scans in syndromic children. The significance of these and the long-term outcomes are still unclear. The true incidence of arachnoid cysts in the general population is also still up for debate. Arachnoid cysts have been reported to be more common in children (2.6%) than in adults (1.4%).[4,5] Paediatric intracranial arachnoid cysts do result in invoking a lot of anxiety to parents, families and primary care teams, with the most common concern being the possibility of intracystic haemorrhage, association with migraines/headaches and concerns on future neuro cognitive and behavioural development.

MATERIALS AND METHODS

We carried out a retrospective review of all cases of paediatric arachnoid cysts diagnosed at the Royal Hospital for Children and Young People (RHCYP) in Edinburgh, Scotland. The period under review was from January 2015 to September 2020. The charts were identified based on radiological reports on all cranial

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ultrasounds, CT and MRI scans. The inclusion criteria included patients aged 0 to 18 years, radiological confirmed arachnoid cyst as reported by a paediatric neuroradiologist and availability of medical records with detail clinical history, examination findings and evidence of clinical follow up after initial presentation. The data collected included age at diagnosis, gender, location of arachnoid cysts, reason for investigation and management plan. Ethical approval was obtained for this retrospective audit study (Table 1).

RESULTS

Initially 285 patient records were retrieved from radiology reports, 98 records were identified as duplicates and a further 20 patient records were discarded due to incomplete data / follow up. A total of 167 records were therefore available for analysis.

In terms of gender, there were more males than females with diagnosed arachnoid cysts (61% vs 39%) (Figure 1). The predominant location of the cysts was the middle cranial fossa followed closely by the posterior cranial fossa (Figures 2 and 4). With regards to laterality, most cysts were located on the left side (47.3%), followed by a midline location in 26.9% (Figures 3 and 4).

DISCUSSION

Arachnoid cysts are a common cause of referrals to the paediatric neurosurgeon. Once diagnosed, they are often a cause for anxiety for patients and their families. The true incidence of their occurrence in the general population is unknown. Various estimates ranging from 0.3% to 1.4% in children and up to 2.6% in adults have been reported.[4-8] This is especially challenging as they are a radiological diagnosis, and most are found while investigating for an underlying neurologic concern. The true prevalence in the general population will therefore continue to be elusive.

Though largely labelled as congenital anomalies,[9] the exact aetiology of arachnoid cysts is still controversial. The evolution and natural history of lesions antenatally is also scant and few intrauterine studies are available that demonstrate actual evolution. Grossman *et al.* in were able to demonstrate that in antenatally diagnosed arachnoid cysts, subsequent scans showed regression in 10% and progression in 10%. The majority (80%) as

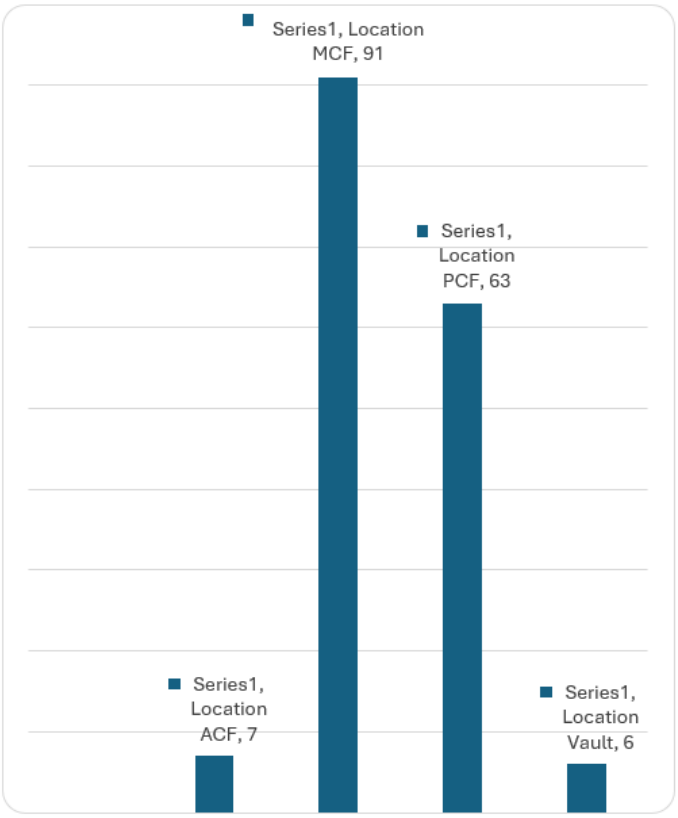

Figure 2: Location of arachnoid cysts in intracranial compartment.

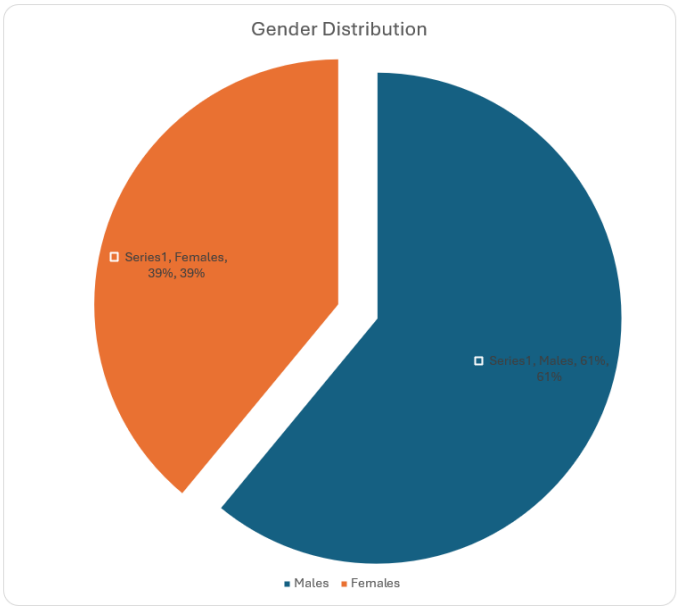

Figure 1: Gender distribution of patients.

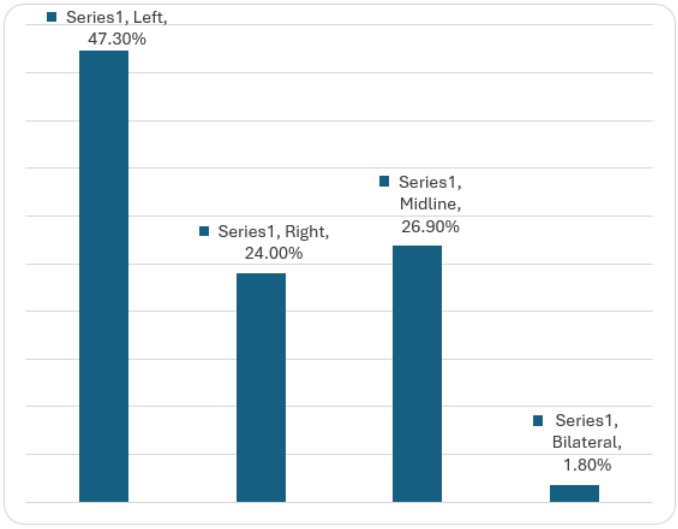

Figure 3: Lateralization of arachnoid cysts.

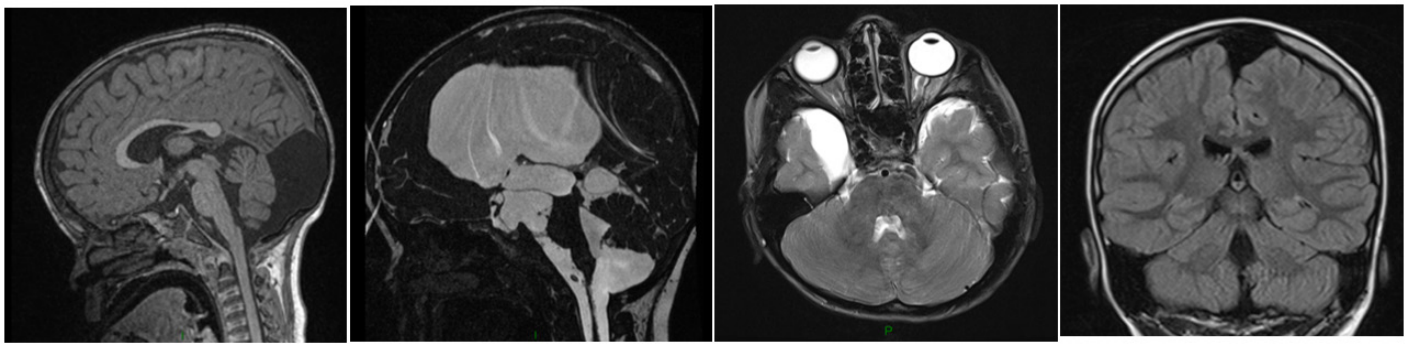

Figure 4: Various locations of arachnoid cysts a) Posterior fossa, b) suprasellar, quadrigerminal and posterior fossa, c) temporal fossa and d) convexity/ parasigital.

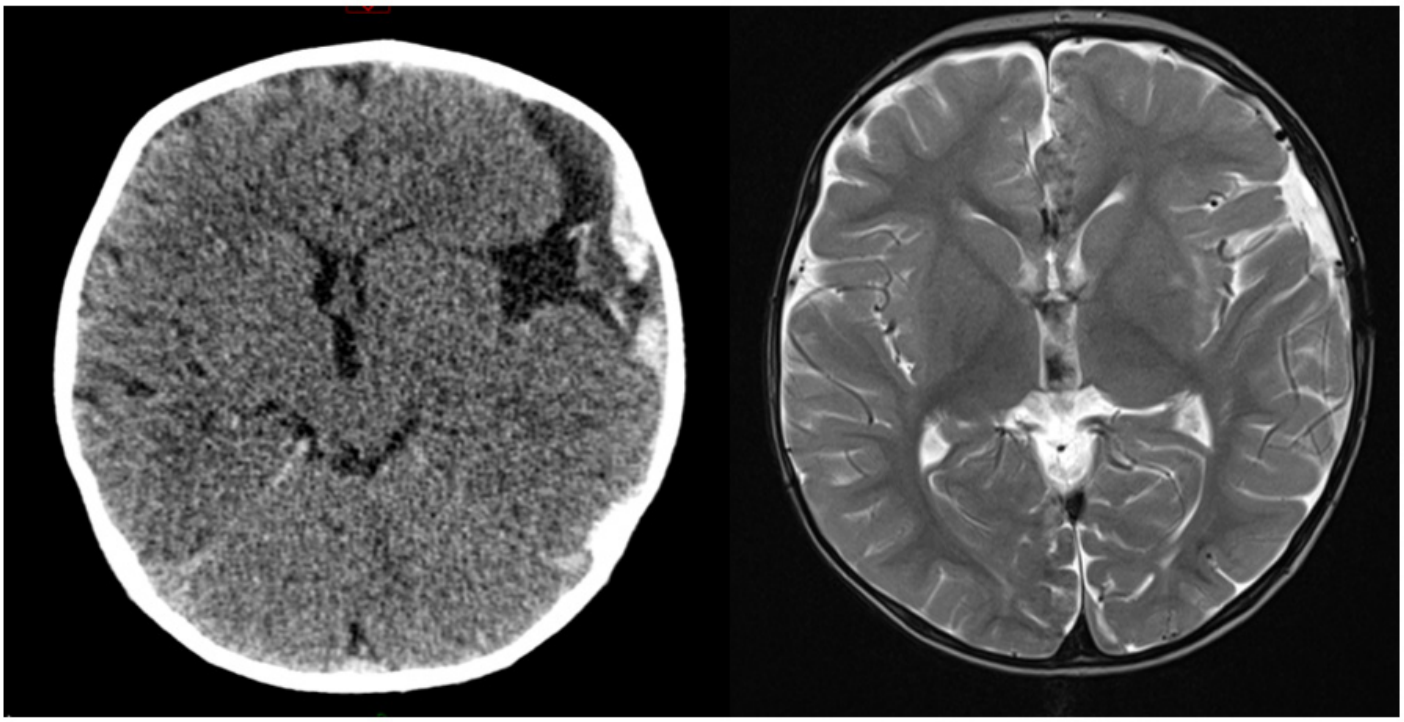

Figure 5: CT scan demonstrating intracystic haemorrhage and ipsilateral suleal effacement (left) and 6 month post operative MRI scan showing regression of arachnoid cyst.

expected were unchanged. There have been reports of post-natal development of arachnoid cysts as well i.e. not visible in antenatal scans but subsequently diagnosed in post-natal scans.[10] In our study, none of the arachnoid cysts were diagnosed prenatally. The earliest diagnosis in our case was at birth (day one of life) and the oldest at 17 yrs. This compares well with most studies showing post-natal rather than antenatal diagnosis.

The anatomic location of the arachnoid cysts are often of medical significance. This is because most cysts are associated with local mass effect. The symptoms are therefore influenced by the function of the surrounding cortex. As such varied symptoms may include weakness/paraparesis (motor cortex), sensory changes (sensory cortex) etc. Hydrocephalus may also result from impaired CSF flow due to quadrigeminal, suprasellar and posterior fossa arachnoid cysts. These often obstruct CSF flow in association with seizures have also be noted be related to frontal and middle cranial fossa arachnoid cysts. Various studies have reported varying incidences by location. In our series, the most common location was in the middle cranial fossa (54.5%) followed by the posterior cranial fossa at 37.7%. This is significantly different from the antenatal series by Grossman *et al.*[11] that demonstrated an interhemispheric location at 85.6% and middle cranial fossa location in a paltry 8.6%. It is possible that many of the interhemispheric arachnoid cysts regress before birth accounting for their low numbers in most post-natal studies.

An interesting finding in our series was with regard to the lateralisation of location of the arachnoid cysts. A majority were left sided in location (47.3%) compared to right sided cysts that comprised 24% of patients. This is worth noting as a majority of the population are left side dominant and hence injuries or neurologic deterioration due to the left sided cysts may result in significant neurologic sequelae. The significant number of

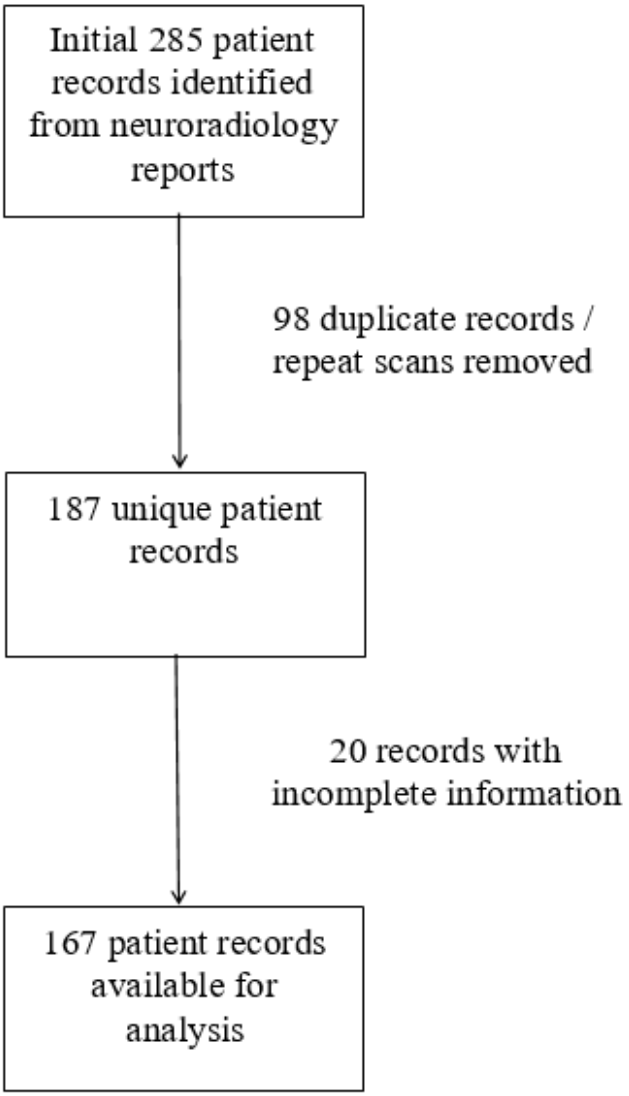

Table 1: Study outline.

midline arachnoid cysts 26.9% may also result in hydrocephalus due to obstruction of CSF flow.

Several syndromes have been reported to be associated with increased incidence of intracranial arachnoid cysts[12] such as:

- Down syndrome
- Mucopolysaccharidosis
- Schizencephaly
- Neurofibromatosis
- Autosomal dominant polycystic kidney disease
- Acrocallosal syndrome
- Aicardi syndrome

In our series, 16 patients (9.6%) had associated syndromic diagnoses such as Soto's syndrome or Downs syndrome. Other common causes for investigation with neuroimaging that led to a diagnosis of arachnoid cysts were seizures in 18 patients (10.8%), headaches in 13 patients (7.8%) and an abnormal head shape. Various genetic mutations or syndromic cases may be associated with arachnoid cysts. It is unclear whether this represents a causality/association or just that patients with a suspected syndromic cause often get more imaging. Further studies in this area would be beneficial.

Most cases of arachnoid cysts can be managed non-surgically. In our series, 157 patients (94%) were managed conservatively. None of these patients ever needed surgical intervention later on despite prolonged follow up. Of the 10 patients (6%) who needed surgery, 5 patients had features of raised intracranial pressure due to either intracystic haemorrhage or hydrocephalus Figure 5. This is in keeping with similar reports in neurosurgical literature.[13,14] Out of these, 2 patients had craniotomy and evacuation of intracystic haemorrhage, 5 patients had endoscopic fenestration, while 3 patients had shunt insertion. The surgical intervention was at the discretion of the treating surgeon[15] confirmed identical quality of life years no matter the method of surgical intervention used.

CONCLUSION

In conclusion, arachnoid cysts still remains a diagnostic and management challenge. Majority of the cases are often incidental and can be managed without operative intervention. Once the diagnosis is made, it is important to have a detailed discussion with the parents of these children to explain long term implications, participation in contact sports and the need for clinical and radiological follow-up if the children are symptomatic.

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CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

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