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## RESEARCH ARTICLE

# ACCES SCALE A NEW INSTRUMENT FOR ASSESSING CEREBRAL ARACHNOID CYSTS IN CHILDREN

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**ABSTRACT**

Arachnoid cerebral cyst in children are a pathology which induces fear, anxiety and false perceptions in the children's affected family. Monitoring arachnoid cysts on long term can prevent the evolution of the child to a lower quality of life and the aggravation of the condition in some cases. Wetake in study 83 children which were diagnosed with computer tomography or magnetic resonance with arachnoidcerebral cystsin the period 2014-2016 and we applied on them a new grading scale for arachnoid cysts, arachnoid cerebral cysts children evaluating scale ( ACCES scale). This scale contains four items: 1) the clinic involvement 0-3, 2) compression phenomena 0-3, 3) presence or absence of seizures, 4) cognition (memory, thinking), language or behavioural disturbances 0-3. So by far the child is evaluated and we can recognise the most severe cases and make intervention plan. There were evaluated 83 children with imagistic discovered arachnoid cerebral cysts. A male predominance was observed 59 boys and 24 girls. The age when the cyst were discovered was at 10 years old, by far the most. Cyst size was more often in the greatest diameter of 20-30 mm (31 children). There were children with one or more arachnoid cysts. Also there were children with associated malformations. In our series the highest ACCES score was 6, but a score higher than 4 influences the daily life of the patient. The proposed method is of great clinic utility, is not invasive and brings not new costs for patient investigation.



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## INTRODUCTION

Arachnoid cyst in children induces fear and anxiety in the families having children diagnosed with this pathology. From neuroembryological point of view arachnoid cysts are due to the expansion of the roof plate of the brain vesicles or the cyst can be formed from extra axial structures like the arachnoid membrane or migrating ependymal cells (Utsunomiya H1, Yamashita *et al*., 2006).

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Reverse as they appear, arachnoid cyst can disappear in very rare cases and this happens when a cyst rupture is produced followed by subdural effusion and then resorption (Toshiki Mori, Masahito Fujimoto & Satoshi Ueda *et al*., 1995). Some authors describe a continuum of epithelium lined cysts when they are located in the pituitary region like craniopharyngioma (CR), Rathke's cleft cyst (RCC) and intrasellar arachnoid cyst (AC) and so the difference between them sometimes is hard to made preoperatively (Jennifer L., Shin Sylvia and Shereen Ezzat *et al*., 1999). Sometimes arachnoid cysts are incidental discovered and in some pathologies like uncomplicated migraine or defined clinical headache syndrome neuroimaging

is not always recommended but just when these entities are very well defined (Lewis DW, Dorbad D., 2000). In general adult population the prevalence of arachnoid cyst is more in the middle cranial fossa (65,2%) and more frequent in means that in females and have more an unilateral distribution in special to the left (Wester Knut.1999). The arachnoid membrane can be destroyed in time by the growing of the arachnoid cyst (Zhen Tan, Yongxin Li and Qian Chen *et al*., 2015). When the cyst is located in the left frontal fossa near the cyst in the left temporal lobe can exist dysfunction of the left temporal lobe demonstrated with SPECT imaging casing delayed language and learning difficulty (Horiguchi T, Takeshita K., 2000). In some cases the temporosylvian cysts can lead to the extreme pneumatisation of the sphenoid and frontal sinuses without mass effect (Zara G, Ponza I, Citton V, Manara R., 2010). In fetuses cisterna magna has different sizes growing with the fetus, but assessing cisterna magna with ultrasound at 11-13 weeks of gestationusing mid-sagital viewcan bring data about developing arachnoid cysts in this age period (Garcia-Posada et al., 2013).

But in some cases even if chronic local intracranial pressure exists neurologic symptoms or intellectual disabilities are not the rule (Ulmer et al., 2002). Arachnoid cyst can be classified in 3 types which are following afterthatdifferent types of neurosurgical procedures. So are complete communicating, incomplete communicating, and noncommunicating, arachnid cerebral cysts (Zhen Tan, Yongxin Li and Qian Chen *et al.*, 2015). In this forms with cysternography we can obtain the filling of the cyst in 1 h in complete communicating cysts in 3 hours begin the cyst filling in the incomplete communicating cyst forms and no or slight filling in 24 hours in the noncommunicating cysts (Wang X1, Chen JX, You C, Jiang S.,2012). Suprasellar arachnoid cysts can be classified as follows: SAC1 due to the expansion of the diencefalic leaf of the Lliequist membrane, SAC2 due to themodification of the mesencephalic leaf of the Liliequist membrane and SAC 3 including the asymmetrical forms expanding to the subarachnoid spaces (André A1, Zérah M and Di Rocco F. *et al*., 2016). In the evaluation of the middle fossa arachnoid cyst was used the Cambridge Neuropsychological Test Automated Battery (CANTAB)which is alaptop based neuropsychological test battery to evaluate the cognitive improvementafter decompression surgery (Torgersen J1, Helland C, Flaatten H, Wester, 2010).

In the evaluation of children with arachnoid cysts intelligence test can show discrepancies between verbal IQ and performance IQ from the Wechsler Intelligence Scale for Children-Revised (WISC-R), which can be corrected after surgical intervention (Matsuda W, AkutsuH,&Matsumura A.et al., 2010). Some authors show that arachnoid cysts may not be related to a specific seizure type and also may not be in accordance to a specific EEG focus (Yalçin AD, Oncel C, Forta H. *et al*., 2002). Epilepsy is indicated for cyst surgery in 25% cases of the operated cysts together with other pathologies for which operation is performed likeintractable headaches (45%), increasing cyst size (21%) and hemiparesis (8%) (Michael L. Levy,Michael Wang,Hal Meltzer *et al*. 2003). Rare neurologic pathology related to arachnoid cysts: A arachnoid cyst of the Meckel cave gave a neurovascular compression cynsrome resulting in a trigeminal neuralgia and patient improvement was obtained with conservative treatment. (Grasso G, Passalacqua M and Tomasello F. *et al*., 2014).

In the literature is described a case of an adolescent with muscial hallucinations related to an large arachnoid cyst and they included sirenes sounds of dogs barking and children crying, hyms, the voice of her sister. She has also progressive deafness, so the hallucination were organic and imaging revealed the cyst (Griffiths, 2000). In the middle fossa arachnoid cysts one option is the cystocistern fenestration (Li *et al*., 2013) preoperatively and postoperatatively visual analogue scales were applied for patients with arachnoid and dizziness due to arachnoid cysts showing that with low risk of complications after surgery life –quality improved (Mørkve *et al*., 2016). In the surgical treatment of arachnoid cysts in children each case is evaluated and individualized treatment is applied andsurgical methods can be open craniotomy for cyst excision or fenestration, endoscopic fenestration, and cystoperitoneal shunting (Ali *et al*., 2014). In the suprasellar arachnoid cysts ventriculocystocisternostomy is a better method than ventriculocystostomy (Ozek, 2013). In the operations of the quadrigeminal cerebral arachnoid cysts reconstruction of the cerebrospinal fluid circulation is important and for this nowadays is used the frameless navigation (Hu *et al*., 2011). In the noncommunicating arachnoid cysts with definite neurological findings surgery is needed and neuroendoscopic cystic fenestration is a minimally invasive method (Huang *et al*., 2007). Arachnoid cysts are inducing anxiety and anxiety levels of this patientsare higher than in general population and so quality of life is influenced in persons having one or more arachnoid cysts (Spansdahl, 2007). After surgical treatment some author describe clinical improvement of patients just in 60 percent of them (Zhen Tan, *et al*., 2015). After arachnoid cyst chronicsubdural hematoma andhygroma can develop and reverse when in patients this pathologies were investigated in 2,43% of them arachnoid cyst of the middle cranial fossa were found (ParschCathrin *et al*., 1997). So arachnoid cysts have a different pathophysiology, different anatomic and morphologic appearance, a large variety of symptoms and even if different authors have sometimes divergent points of view their presence must be carefully examined and assessed, many pathologies are nowadays not fully known and many researches need to be made in the genetic field there where arachnoid cysts are involved.

## MATERIALS AND METHODS

In children arachnoid cysts are not very well known and their effects tend to be sometimes mysterious and parents don't find always the answer that they expect form the caregivers. This study want to offer new instruments for assessing arachnoid cysts, which can help to recognise the degree of organic involvement in termsof the imagistic appearance and the subjective perception of the presence of the cyst by the children as also by the families of the little patients. Evolution of these cysts varies, for example the prognostic of the largecysts of the temporal fossa is much worse than those of little dimensionsof the posterior fossa.Early detection of arachnoid cysts through imaging investigation methods allows the development of models to prevent further adverse unfavourable evolution, allowing a better quality of life for the patient.In the last years that is why lately there are more and more new researches that approach intracranial arachnoid cysts from different point of views, but there is still no grading system that allows them to be graded on a scale of gravity. The present study was made in the neurology service of the Emergency Hospital for Children "SfântulIoan"Galați and the children were investigated in our hospital with MRI and EEG

in the period 2014-2016. The inclusion criteria in the study were thepresence of the cerebral arachnoid cyst on MRI despite of the dimensions, localisation, form or age of diagnosis.The difference with MRI and EEG between congenital or gained arachnoid cyst is quite impossible and sometimes this has no importance for the patients well-being.We imagined a new instrument to determine the gravity of thepatients damage which we called Arachnoid Cyst Children Evaluating Scale or ACCES Scale.We took four items to be included in the scale:

**The degree of clinic involvement 0-3: 0-no clinic involvement**

- low clinic involvement
- impairment in some daily activities
- impossibility of daily activities andserious
- impairment in daily life

**Compression signs 0-3: 0-nocompression signs**

- imagistic observed compression signs
- imagistic signs, vomiting, or seizures
- imagistic signs, headache vomiting and seizures which can rapidaggravate child's condition

**Convulsions 0-3: 0-no convulsions**

- one episode of convulsions
- two or more episodes of convulsions or epilepsy in treatment with controlled seizures more rare than 1 seizure /month
- no seizure control, seizures more often than 1 seizure/month

Cognitive impairment (memory, thinking) the language impairment or behavioural disturbances0-3: 0-no cognitive impairment

- mild signs of cognitive impairment
- medium cognitive involvement
- severe cognitive impairment

There will be 4 severity degrees:

- Low degree arachnoid cyst scale:0-2
- Medium degree arachnoid cyst scale: 3-5
- Severe degree arachnoid cyst scale: 6-9
- Very severe degree arachnoid cyst scale: 10-12

The study also proposes an algorithm for the exploration and treatment of children who have been found to have arachnoid cysts.

## RESULTS

We took in study 83 children.

| Gender | Boys | Girls |
|---|---|---|
| Total patients | 59 | 24 |

Figure 1 shows the boys/girls repartition: column at 1 are the boysandcolumn 2 are the girls.

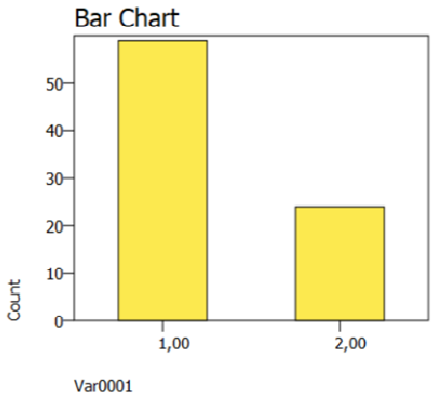


**Figure 1. Bar charts of girls and boys repartition**

So in the arachnoid cyst repartition between genders we saw the predominance of the arachnoid cysts in boys, so it was described in the specific literature. On different age groups arachnoid cysts have a predominance at the age>5-10 years (29 children), but have a greater prevalence at>1-5 years (19 children) and respectively>10-15 years (20 children). The most reduced frequency of cases is at 0-1 year. Table 2 shows the age repartition and Figure 2 alsothe presence of arachnoid cysts at different ages, while Figure 3 shows the standard deviation and the histogram of this age grouping list.

**Table 2. Age groups repartition of the arachnoid cysts in the children age groups**

| AGE GROUPS | 0-1 | >1-5 | >5-10 | >10-15 | >15-18 |
|---|---|---|---|---|---|
| TOTAL | 7 | 19 | 29 | 20 | 8 |

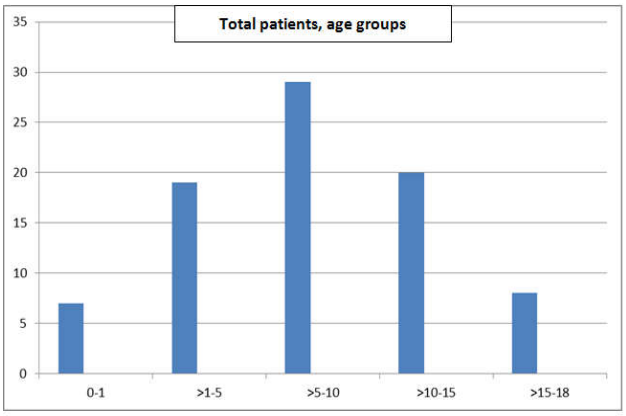


**Fig 2. Age repartition groups on the patients with arachnoid cysts**

The dimensions of the arachnoid cyst we took after the greatest value in one axis, so we have cysts with dimensions<10 mm (10 children), > 10-20 mm (31 children), > 20-30 mm (11 children), > 30-40 mm (19 children), > 40-50 mm (10 children)and over 50 mm (2 children). The predominance is that of the cysts with little dimensions and the most rare are the cysts with dimensions greater than 50 mm. The great arachnoid cysts were found in the temporal fossa so they are described in the literature.Figure 4 shows a great temporal fossa arachnoid cyst from our series. The arachnoid cyst can fill the whole temporal fossa and then appear also compression signs. In such a case the skull can suffer deformation and the head is asymmetric. In these cases the children are very affected being in the school the victims of bullying (teasing, marginalization in school) and their school performance often decreases on psychological mechanisms and not necessarily

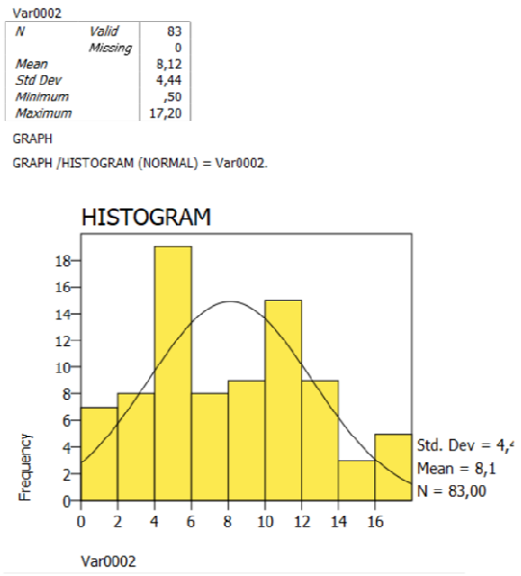


**Fig 3. Age repartition histogram**

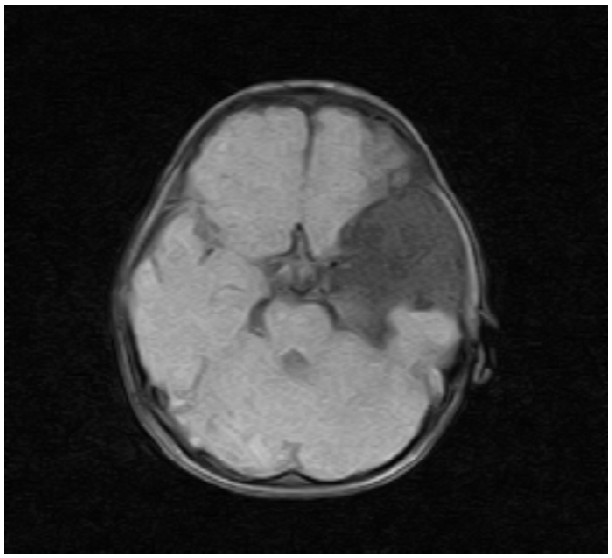

**Figure 4. Left temporal fossa arachnoid cyst**

due to volume or compression on the underlying temporal structures. We can have children with one arachnoid cyst, children with 2 arachnoid cysts, or children with 3 arachnoid cysts according to the number of arachnoid cysts. There are 72 children with one arachnid cyst, 8 children with 2 arachnoid cysts, 3 children with 3 arachnid cysts. Cyst localisation was found in our series:interhemisferic,frontoparietal left,temporal anteriorright,temporal right,temporalleftwith mass effect, temporal left of millimetre,suprasellar, behind the cerebellum, posterior fossa arachnoid cyst, pontocerebelos angle, Infravermian, occipital,temporopolar bilateral,frontobasal right, several subtentorial cysts (3) -1 case, supra andand infratentoral-3cases.

**Table 3. Total patients number of cysts**

| NUMBER OF CYSTS | 1 | 2 | 3 |
|---|---|---|---|
| TOTAL PACIENTI | 72 | 8 | 3 |

Figure 5.Number of patients with one or more arachnoid cysts
The most important symptoms for which the children presented in or service were: headache (24 children), headache with cranial hypertension -2, migraine-3, hemiparesis 3,

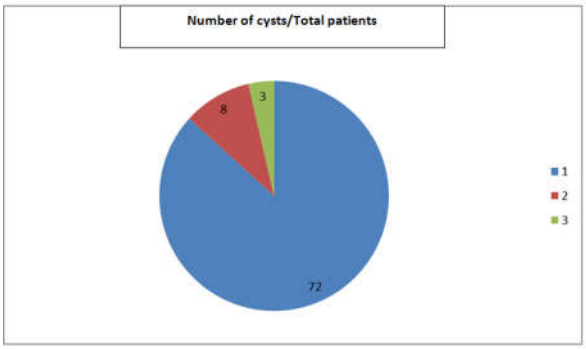


**Figure 5. Number of patients with one or more arachnoid cysts**

triparesis-1, tetraplegia 2, delay of language with depigmentation stain, isolated language disorders, hydrocephaly, ethmoidomaxillary sinusitis, hypersomnia, cerebellar syndrome, paroxistic phenomena, febrile convulsions, vertigo, Bells Palsy, transitory deficit of a hemibody, partial epileptic seizures, afebrile tonic clonic seizures, amnesia, tics, head trauma with loss of consciousness, epileptic encephalopathy, ethmoidomaxilar sinusitis,nistagmus, hypogonadotrop hypogonadism. Not always the symptoms for which the children present is pathology so for example is it after loss of consciousness, so we cannot affirm that he will develop epilepsy even if the EEG pattern is modified and a arachnoid cyst and a seizure with loss of consciousness is present.

As associated pathology we found more elements which can raise the ACCES score, so for these malformations we will give 1-2 points and are included in the clinic spectrum segment. To this malformations can assigned a nowadays unknown mutation or anassociation of symptoms not described until yet by other authors.An important observation to be made is that we won't quote the left neurologicallateralisation in this children, considering it physiological the same as in other children. The parents tend to put the left neurological lateralisation in the symptoms cohort of the arachnoid cerebral cyst in their child considering it most when the cyst is on the right side. The parent will be advised in this cases not to change the child's lateralisation because of the negative effects of the „reversal" of the child's hands.

Table 4 and Figure 6 shows the proportion of children found with compression signs

**Table 4 Number of children with compression signs**

| SIGNS OF COMPRESSION | YES | NO |
|---|---|---|
| TOTAL PATIENTS | 7 | 76 |

The ACCES score of the patients is shown in the table 5 and Figure 7. So this score shows the repartition of the most severe cases.

**Table 5 The ACCES score of the evaluated children**

| ACCES SCORE | 0 | 1 | 2 | 3 | 4 | 5 | 6 |
|---|---|---|---|---|---|---|---|
| TOTAL PATIENTS | 17 | 34 | 12 | 12 | 2 | 2 | 4 |

Figure 7. The repartition of patients on degrees on severity on ACCES score

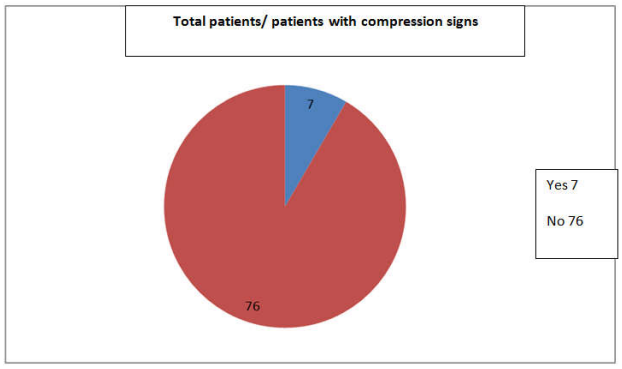


**Figure 6. Number of patients with compression signs**

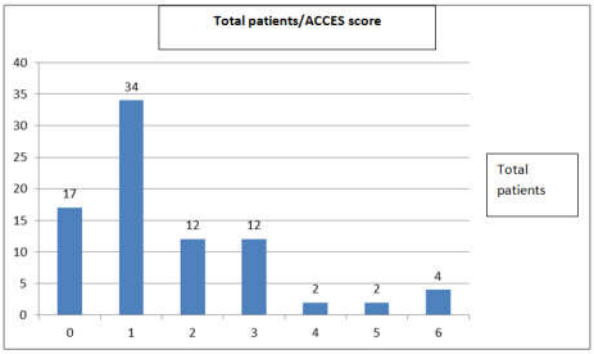


**Figure 7. The repartition of patients on degrees on severity on ACCES score**

The most children have not major clinical implications due to the arachnoid cysts, because their score is just of 0 and 1. In the 0 group the symptoms which bring the child to the doctor were incidental, but the child was explored and the arachnoid cyst was found. That is not meaning that the brain of the child in later life cannot develop other pathologies so the child is hold under surveillance. In the score 1 group symptoms were also rare or very mild. As the score gets higher there are few children with this daily life impairment but their number must not be neglected. We can see that children with severe implication were found in our group.

## Conclusion

The method is of great clinic utility and is not invasive and brings not new costs for the patient's investigation. The ACCES scale can be appliednot only by the neurosurgeon or the neurologist but also by the family doctor which known very well the case documentation, the family of the child with its hole history, the child's comorbidity, the living environment, the perception about the symptoms from the child as also from the relatives depending on the microclimate and the sociocultural environment.This instrument is also a common language between specialists and so the evaluation of the child is more easier, and it's more easier to plan rehabilitation and prevention for the child as also to plan prevention for a greater population form a district hold under surveillance for this pathology. Cerebral arachnoid cyst must not create panic to the child and the parent but they must be managed with caution and a correct appreciation of them is beneficial for the child's neurologic and cognitive evolution.

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