



## SCIENTIFIC LETTERS

### Quality of life heart-disease children who have suffered from an arterial ischaemic stroke<sup>☆,☆☆</sup>



### Calidad de vida de niños cardiopatas que han sufrido un ictus arterial isquémico

Dear Editor:

Arterial ischaemic stroke is infrequent in the paediatric population. Between 50% and 80% of these patients develop sequelae that affect different areas.<sup>1</sup> Heart disease is one of the most frequent causes of stroke in children and accounts for nearly one third of cases.<sup>2</sup>

The term health-related quality of life (HRQoL) refers to the impact of a disease and its treatment in the physical, psychological and social wellbeing of a patient as perceived by the patient. In recent years, there has been a growing interest in researching the quality of life of children that have experienced a stroke.<sup>3,4</sup>

Our objective was to assess HRQoL in a sample of children with heart disease who had suffered an arterial ischaemic stroke.

We recruited patients from a larger case series that analysed the outcomes of children with heart disease who had experienced an arterial ischaemic stroke between January 2000 and December 2014.<sup>5</sup> Patient outcomes were evaluated by means of the Paediatric Stroke Outcome Measure (PSOM) and the modified Rankin scale (mRS) at least 6 months after the stroke. At the time, HRQoL in patients aged more than 5 years was assessed by means of the generic core scales of the Paediatric Quality of Life Inventory (PedsQL<sup>®</sup>) version 4, for which we obtained authorisation. The PedsQL<sup>®</sup> comprises 15 items, 5 on physical functioning, 4 on emotional functioning, 3 on social functioning and 3 on school functioning, in

addition to a total scale score. The score can range between 0 and 100, with 0 representing the least quality of life and 100 the most.

In the statistical analysis of the data, we used the one-sample *t* test to compare the PedsQL<sup>®</sup> scores of our patients with those of healthy controls and children with other diseases reported in the reviewed literature.

Thirty-four children aged more than 5 years completed the quality of life questionnaire. Table 1 summarises the demographic characteristics of the patients, as well as the clinical features of their heart disease and stroke. The mean time elapsed between the stroke and the assessment was  $6.9 \pm 4.1$  years.

The PedsQL<sup>®</sup> total scale score was lower in patients that had poorer outcomes according to the PSOM ( $60.11 \pm 10.95$  vs.  $69.44 \pm 14.16$ ;  $P = .038$ ) and the mRS ( $52.50 \pm 7.32$  vs.  $66.10 \pm 13.09$ ;  $P = .052$ ), especially in the physical functioning subscale.

Patients with epilepsy had significantly lower scores in the physical functioning subscale of the PedsQL<sup>®</sup> ( $51.58 \pm 20.36$  vs.  $74.18 \pm 16.77$ ;  $P = .001$ ). We also found differences in the scores of patients with clinically significant hemiparesis in the physical functioning subscale ( $55.38 \pm 20.01$  vs.  $72.90 \pm 18.92$ ;  $P = .015$ ) and the total scale score of the PedsQL<sup>®</sup> ( $58.62 \pm 11.48$  vs.  $68.14 \pm 13.18$ ;  $P = .039$ ). Large size of the infarct was associated with significantly lower scores in the physical functioning PedsQL<sup>®</sup> ( $55.13 \pm 20.37$  vs.  $70.23 \pm 19.74$ ;  $P = .04$ ) due to greater motor deficits.

The scores in our patients were significantly lower than those of healthy controls reported in the literature<sup>6</sup> (Table 2).

Furthermore, our patients had a poorer HRQoL compared to children with heart disease that had not had a stroke except in the area of emotional functioning. The PedsQL<sup>®</sup> scores of our patients were significantly lower than those of children with other chronic diseases (diabetes), and similar to those of children with chronic asthma (Table 2) except in the school functioning subscale (worse in our patients).<sup>6</sup> This is noteworthy, as children with chronic asthma are a population described in the literature as having poor HRQoL.

The poor HRQoL outcomes in our patients may be explained not only by the stroke, but also by the severity of heart disease. Thus, patients with more severe disease, such as hypoplastic left heart syndrome, scored significantly lower in the school and social subscales.

Comparing the PedsQL<sup>®</sup> scores of our patients to those of other children in the literature, we found that they scored

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<sup>☆☆</sup> Previous presentation: the results of this study were presented at the Congreso de la Sociedad Española de Neurología Pediátrica (SENEP), held between May 19 and 21, 2016 in Toledo, Spain, as part of the oral presentation "Pronóstico del ictus arterial isquémico en niños con cardiopatía", which received one of the awards to the best SENEP2016 presentations.

**Table 1** Characteristics of heart disease and stroke.

Demographic and heart disease characteristics		Stroke characteristics	
<i>Age at stroke, months; median (IQR)</i>	32.3 (5.0–67.4)	<i>Manifestations at onset:</i>	
<i>Male sex</i>	22/34 (65%)	Motor deficit	23/34 (68%)
<i>Left ventricular hypoplasia</i>	10/34 (29%)	Convulsive seizures	11/34 (32%)
<i>Congenital heart defect</i>	30/34 (88%)	Sensory deficit	4/34 (12%)
<i>Other associated cardiovascular factors:</i>		Aphasia	7/34 (21%)
Left heart valve disease	7/34 (20%)	Asymptomatic	3/34 (9%)
Arrhythmia	11/34 (32%)	<i>Radiologic findings:</i>	
R-L shunt	21/34 (62%)	Multiple stroke	12/34 (35%)
Dilated cardiomyopathy	6/34 (18%)	Bilateral stroke	10/34 (29%)
Thrombus in left chamber	2/34 (6%)	Unilateral stroke: left/right	33/67%
Thrombus in artery/vein	2/34 (6%)		
Endocardial device	10/34 (29%)		
<i>Stroke associated in time with a procedure:</i>			
Diagnostic cardiac catheterization 72 h before	0/34 (0%)	Circulation: anterior/posterior/both	79/6/15%
Therapeutic cardiac catheterization 72 h before	7/34 (21%)	Stroke size: lacunar/medium/large	6/50/44%
Heart surgery 72 h before	5/34 (15%)	Haemorrhage	10/34 (29%)
Heart surgery 72 h-1 month before	14/34 (41%)		
ECMO or ventricular assist device	5/34 (15%)	<i>Stroke recurrence</i>	2/34 (6%)

**Table 2** Quality of life scale scores (PedsQL<sup>®</sup>) in our patients compared to those of healthy children and children with other chronic diseases.

PedsQL <sup>®</sup> scale	Children with heart disease and stroke (our series)	Healthy children <sup>6</sup>	Children with heart disease <sup>6</sup>	Children with asthma <sup>6</sup>	Children with diabetes <sup>6</sup>
<i>N</i>	34	5480	344	157	300
Physical functioning	66.2 ± 20.9	87.5 ± 13.5; <i>P</i> < .001	83.1 ± 18.7; <i>P</i> < .001	72.6 ± 18.3; <i>P</i> = .080	85.8 ± 13.3; <i>P</i> < .001
Psychosocial health summary score	63.8 ± 13.3	81.9 ± 14.1; <i>P</i> < .001	77.3 ± 17.2; <i>P</i> < .001	66.8 ± 16.6; <i>P</i> = .170	77.3 ± 14.6; <i>P</i> < .001
Emotional functioning	68.3 ± 17.7	79.3 ± 18.2; <i>P</i> = .001	74.6 ± 20.4; <i>P</i> = .040	64.8 ± 20.2; <i>P</i> = .200	72.3 ± 19.5; <i>P</i> < .001
Social functioning	69.2 ± 19.2	85.2 ± 16.8; <i>P</i> < .001	82.5 ± 20.1; <i>P</i> < .001	73.4 ± 18.6; <i>P</i> = .210	85.6 ± 16.2; <i>P</i> < .001
School functioning	53.5 ± 16.3	81.1 ± 16.5; <i>P</i> < .001	73.0 ± 20.3; <i>P</i> < .001	62.3 ± 20.6; <i>P</i> = .003	74.2 ± 18.0; <i>P</i> < .001
Total scale score	64.5 ± 13.2	83.8 ± 12.7; <i>P</i> < .001	79.4 ± 16.5; <i>P</i> < .001	68.7 ± 15.9; <i>P</i> = .680	80.3 ± 12.8; <i>P</i> < .001

Data expressed as mean ± standard deviation.

lower than child survivors of stroke of any aetiology and similarly to child survivors of stroke with a poor outcome.<sup>4</sup>

The HRQoL of children with heart disease that had suffered a stroke in our series was poorer than that of healthy children and also children that only had a stroke or only had heart disease. Furthermore, patients with unfavourable functional outcomes, with significant hemiparesis or epilepsy, had a poorer quality of life, especially in the area of physical functioning.

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## Jarcho-Levin and Rokitansky syndromes. An exceptional association<sup>☆</sup>



### Síndromes de Jarcho-Levin y Rokitansky. Una excepcional asociación

Dear Editor:

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is characterised by congenital aplasia of the uterus and the upper 2/3 of the vagina in women showing normal development of secondary sexual characteristics and a normal 46, XX karyotype.<sup>1</sup> Its estimated incidence is of 1 in 4000–5000 live female births.<sup>2</sup> Jarcho-Levin syndrome (JLS) or spondylocostal dysostosis is defined by the association of costal and vertebral malformations resulting in a shortened trunk and short stature.<sup>3</sup> It occurs in 1 out of 4000 live births.<sup>4</sup> Here, we present the case of a female patient with a neonatal diagnosis of JLS who sought care for primary amenorrhoea, the evaluation of which revealed Müllerian agenesis.

The patient was a girl aged 15 years with a diagnosis of primary amenorrhoea. The prenatal history was unremarkable. She received a diagnosis of JLS at birth based on the presence of a shortened trunk and thoracolumbar malformations. The patient also had a perimembranous ventricular septal defect, an ostium secundum atrial septal defect and a right inguinal hernia containing the right ovary.

At the time of assessment she had completed puberty (Tanner stage V) with normal female genitalia. We did not observe any signs of hyperandrogenism. A pelvic ultrasound scan, bone age study and measurement of hormone levels were requested for evaluation of the primary amenorrhoea. The ultrasound examination found no evidence of a uterus or ovaries. The bone age was consistent with

the chronological age. The levels of estradiol (92 ng/mL) and gonadotropins (luteinising hormone, 13.1 mIU/mL and follicle-stimulating hormone, 4.6 mIU/mL) were in the normal range. The patient underwent magnetic resonance imaging of the abdomen and pelvis (Fig. 1), which found that the uterus and the upper third of the vagina were absent. The ovaries were in the normal location, with detection of a cyst in the right one. The morphology and position of the kidneys were normal.



**Figure 1** Pelvic MRI. Sagittal T2-weighted image showing an ovarian cyst against the superior portion of the bladder, and the posterior wall of the bladder adjoining the anterior wall of the rectum due to the absence of the uterus and the upper third of the vagina.

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