

Clinical Spectrum of Cerebral Palsy in South Jordan: Analysis of 122 Cases

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Abstract

Background: The clinical spectrum and associated factors of cerebral palsy (CP) may differ between developing and developed countries.

Aim: To evaluate some predisposing factors, clinical spectrum, and some associated problems of cerebral palsy in children.

Setting and Design: It is a retrospective study where data were extracted from patients file record which contains extensive historical and clinical data in a center for early diagnosis of childhood disabilities.

Patients and Methods: One hundred and twenty two children with ages ranging from 7 months to 17 years were reviewed in a 32-month period between September 2007 and April 2010. A simple statistical analysis was used for a percentage calculation.

Results and Conclusions: A spastic type was predominant (82.7%), with a quadriplegic subtype being the most common (34.4%). The other types were choreoathetoid (8.2%), mixed type (6.6%) and ataxic (2.5%) being the least. Speech delay was the most common associated problem (71.3 %) followed by mental retardation (61.5%), seizures (35.2%), hearing problems (26.2), and autism (4.9%) being the least. The clinical spectrum of CP in Jordan may differ from that reported in Western countries. Prospective studies are needed to evaluate the clinical spectrum and predisposing factors in Jordan.

Keywords: Cerebral palsy, clinical spectrum, south Jordan.

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Introduction

Cerebral palsy is one of the most common and costly chronic disorders. It affects up to 0.02% of all live births, and it occurs in all races. Cerebral palsy was defined as a group of non-progressive permanent disorders of movement and posture that occur following damage to the developing fetal or infant brain. It is often

accompanied by other neurodevelopmental disorders.¹

Reports about the clinical spectrum, predisposing factors and complications are coming from developing countries although many doctors working in this field feel that there is a difference in epidemiology between the reports and what is seen in clinical practice.

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The aim of this study is to evaluate some predisposing factors, clinical spectrum, and some associated problems of cerebral palsy (CP) in children and to compare our figures with national^{2,3} and international studies.

Material and Methods

This study included 122 children seen in the early diagnostic centre which receives all types of children with disabilities. Each child was fully evaluated and a special case record included details about pre- and peri-natal care, family history, developmental milestones, details of clinical examination results of investigations and recommendations. We selected files of CP cases evaluated in this center in a 32-month period between September 2007 and April 2010. All cases were reviewed by the same pediatric neurologist and used the same file records.

Regarding predisposing factors, consanguinity, perinatal complications such as a need for oxygen, resuscitation or NICU admission were considered as one perinatal factor.

Neonatal jaundice was considered if the patient's history indicated phototherapy, exchange transfusion or other treatment modality. Any mild jaundice that did not need medical care or consultation was considered physiological and was excluded.

Regarding associations, epilepsy was considered when it was recurrent and especially in those receiving an antiepileptic treatment.

Cognitive function was assessed by our psychologist when mental retardation was considered clinically or was a concern from the parents although in some it was specifically requested by the education authorities. Speech assessments were done by a speech therapist when needed which was available in our center.

Data were analyzed using a simple statistical analysis for the percentage calculation.

Results

The age of the CP children ranged from seven months to 17 years with a mean age of 6 years and 4 months. Provided we look for clinical spectrum, in this study we included few older children even though in our center we deal with cases up to 18 years.

Among the 122 cases studied, 54.1% were males with a male/female ratio of 1.18 to 1.

Consanguinity of all degrees was found in 53.3% of the cases.

History of perinatal problems was considered if the child needed oxygen, resuscitation or NICU admission. These events were found in 38.5% of the cases, which was considered the most common predisposing factor (see Table 1). In this study, 85.2 % were full term children and 14.8% were preterm. Neonatal jaundice was seen in 36 cases (29.5%). Forty-two (34.4%) of the children were first born, 24 (19.6%) were second, 17 (3.9%) were third and 29 (23.7%) were fourth and another 8.4% were fifth to twelfth in birth order.

Table (1): Predisposing Factors.

<u>Predisposing Factor</u>	<u>(n)</u>	<u>(%)</u>
<i>Consanguinity</i>	65	53.3
<i>Perinatal</i>	47	38.5
<i>N.N jaundice</i>	36	29.5
<i>C/S</i>	33	27
<i>Prematurity</i>	18	14.8

Clinical classification of CP was based upon the type and distribution of motor abnormalities.

Although there was a substantial overlap among the clinical features, the distribution of spastic type was the most common (82.7%), choreoathetoid (8.2%), mixed (6.6%) followed by ataxic (2.5%) being the least (as seen in Table 2). Regarding the spastic type, quadriplegic was the commonest (34.4%), then hemiplegic type (26.2%) and diplegic type (22.1%). Head circumference was measured in all cases, and 48 cases (39.3%) were below or far below the fifth centile for their age.

Table (2): Distributions of Types of Cerebral Palsy.

Type of CP	(n)	(%)
Spastic	101	82.70%
Quadriplegic	42	34.40%
Hemiplegic	32	26.20%
Diplegic	27	22.10%
Choreoathetoid	10	8.20%
Mixed	8	6.60%
Ataxic	3	2.50%

Regarding associated problems (Table 3), speech delay by history or by assessment was the most common (71.3 %) where speech function assessments were done by a specialist speech technician working in our center. Mental retardation was tested with the Stanford Binet intelligence scale, and it presented in 75 of the children (61.5%) with CP. The prevalence of epilepsy in all types of CP in our report was 35.5 %, and its distribution was with quadriplegic CP (40.4%) hemiplegics (31.25%) and in diplegic (25.9%). Hearing impairment was seen in 32 cases (26.2%).

Table (3): Associated Problems.

Associated problems	n	(%)
Speech delay	87	71.30%
Mental retardation	75	61.50%
Seizures	43	35.50%
Hearing impairment	32	26.20%
Autistic behavior	6	4.90%

Complications prevalence also differs according to CP type (Table 4). In the quadriplegic type, there is a higher prevalence for all types of complications. The most striking result was when a child had mental retardation, the chance to have other associations increased to 91.4% for speech, 48% for seizures and 36% for hearing.

Table (4): Prevalence of Complications According to CP Type.

CP type	Seizures	M.R.	Hearing	Speech
Quadriplegic	40.40%	73%	26.10%	76.20%
Hemiplegic	31.25%	46.80%	25%	62.50%
Diplegic	25.90%	37%	14.80%	55.50%

Discussion

Cerebral palsy is the most common neurological disorder seen at neurology clinics.⁴ The etiology of CP is not well understood, and brain lesions are thought to be associated with prenatal, perinatal, or postnatal events of varying causes. Risk factors for CP are multifactorial and can include preterm birth, multiple gestation, intrauterine growth restriction, male sex, low Apgar scores, intrauterine infections, maternal thyroid abnormalities, prenatal strokes, birth asphyxia, maternal methyl mercury exposure, and maternal iodine deficiency.⁵⁻⁷

Consanguinity in all its degrees was found in 53.3% of the children but this is similar to the normal prevalence^{7,8} in our community which is 50 to 65%, which indicates that consanguinity may not be a predisposing factor for developing cerebral palsy. The proportion of paternal parallel first cousins among first-cousin marriages has showed a steady decline from one generation to the next.¹⁰

The role of perinatal complications, particularly birth asphyxia, in the causation of CP has been questioned and asphyxia has been suggested to be a consequence rather than a cause of the process that leads to CP. Seventy-eight percent of children with cerebral palsy did not have birth asphyxia.¹¹ However, we found a history indicative of birth asphyxia in a large number of cases (38.5%), which is similar to the findings of many developing countries.¹² Occurrence of severe birth asphyxia, which is rarely seen in developed countries, continues to be a major problem in many developing countries. Instrumental delivery and caesarian section have been reported to be associated with CP.¹³ However, most of the children in this report were born through normal vaginal delivery (73%).

Multiple pregnancies have been considered as an important prenatal risk factor for CP.¹³ The first four in birth order were 91.6% while only 8.4 % were between the 5th-12th position in the birth order. This means that multiparity has no role as a causative factor for cerebral palsy. This result correlates well with other authors from developing countries.¹⁴

The role of prematurity in our study is clearly limited, given that only 14.8% were born prematurely. Meanwhile, most epidemiologic studies from industrialized countries show a rise in the childhood prevalence of cerebral palsy in recent decades, largely because of the increasing prevalence of children of low and very low birth weight. The only demographic determinant of cerebral palsy's prevalence that is changing rapidly in the United States is the survival of low birth weight and very low birth weight infants. Based on the magnitude of change in the survival of low and very low birth weight infants, it is estimated that the childhood prevalence of cerebral palsy rose about 20% between 1960 and 1986 in the United States. These reports may explain the difference in the type distribution of CP between developed countries and developing countries.

Figures indicate that 27.8% of cases of CP are due to prematurity.¹² CP occurs in 0.2% of live births, but infants born before 28 weeks gestation have a 50-fold elevated risk when compared with infants born at term¹⁵ with prevalence between 6 and 26%.

Neonatal jaundice in our study was present in 29.6 % of the cases but its severity was not measured because historical findings make the value of this finding very limited. In studies from developing¹⁶ countries, the presence of neonatal jaundice as a predisposing factor is 14.4%.

There is no single test to diagnose cerebral palsy. Most of the information leading to the diagnosis of cerebral palsy is generally obtained from a thorough medical history and examination. Once the diagnostic evaluation is complete, further testing may be needed in order to define the specific needs of any individual child.¹⁷

The prevalence of spastic CP cases was 82.7% in our study which is similar to that elsewhere.¹³ The distribution of the clinical subtypes of the spastic cases were very different from the developed countries.¹²

Whereas spastic diplegia is generally the most common type of CP reported from developed countries, in our study spastic quadriplegia was the most commonly seen type in 34.4% of the cases. This was similar to the findings reported by other developing countries¹⁷ as well as Jordanian results. In the Al Ajlouni study, it was 36% and in the AbdelKarim Al-Qudah study, it was even higher reaching up to 41%. This actually needs to be explained so prospective studies need to be encouraged to discover the precise reason why the predisposing factors differ in between different countries.

Also, it is worth mentioning that most cases coming to our center were severe. The most common type of CP was spastic (82.70%) while the next types in frequency were the quadriplegic (34.40%), hemiplegic (26.2%) and the diplegic in (22.1%). In contrast to this, in a study from Boston analyzing 120 cases of CP, the type distribution was 52% had quadriplegia, 31% had diplegia, and 17% had hemiplegia.

Almost all children with CP have at least one additional disability.¹⁸ The most common in this study was a history of speech delay either as a severe delay in the acquisition of the skill or being a persistent problem in 71.3%. A delay in speech development may be a symptom of many disorders, including mental retardation, hearing loss, or psychosocial deprivation which frequently complicates cerebral palsy.

In this study, 61.5% of the cases with CP had mental retardation. Children with spastic quadriplegia had the worst intellectual outcome.

Epilepsy¹⁹ was reported to be seen frequently in CP children in about one third of the cases. It is often severe and difficult to control particularly in children with mental retardation. The presence of cerebral palsy requires differential consideration of the severity of epilepsies. Epilepsy occurs in 35.2% of CP children in this study. Epilepsy was significantly more common in children with CP and mental retardation. Other studies from the Middle Eastern area²⁰ revealed the well-established high frequency of associated neurological deficits.

Regarding autistic behavior, we did not apply any formal test. The diagnosis was done clinically, and the autistic behavior was also observed by our psychologist during the testing of the cognitive function of these children. In the literature, almost nothing is mentioned regarding motor disabilities and autistic disorder.²¹ In this study, all autistic behavior cases were seen in mentally retarded children so possibly the link of autism is more with mental retardation than with CP, provided that the last is a motor disorder.

It was very striking that the presence of mental retardation correlated with the severity of the condition.

In conclusion, the spectrum of CP in Jordan differs from that seen in Western countries.

Perinatal factors are still an important predisposing factor for CP. Prospective studies are needed to evaluate these predisposing factors.

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الشلل الدماغي في جنوب الاردن تحليل ١٢٢ حالة

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الملخص

يختلف الطيف السريري والعوامل المرتبطة بالشلل الدماغي بين الدول النامية والمتقدمة. الهدف من هذه الدراسة هو تقييم بعض العوامل المهيئة، الطيف السريري، وبعض المشاكل المرتبطة بالشلل الدماغي عند الاطفال. وهذه دراسة استيعادية حيث استخلصت المعطيات من ملف سجل المرضى الذي يحتوي على بيانات سيره مرضية وسريية واسعة النطاق في مركز للكشف المبكر عن الإعاقة في مرحله الطفولة. جرى استعراض ١٢٢ حالة تتراوح اعمارهم بين ٧ شهور الى ١٧ سنة في فترة ٣٢ شهرا بين سبتمبر ٢٠٠٧ الى ابريل ٢٠١٠ وتم عمل تحليل احصائي بسيط باستخدام النسب المؤيه وكانت النتائج والاستنتاجات كالاتي:

- النوع التصليبي هو السائد بنسبه ٨٢,٧% الرباعي الاكثر شيوعا ٣٤,٤%
 - الانواع الاخرى الدودي ٨,٢% المختلط ٦,٦% الرنخي ٢,٥% وهو الاقل
- اما العوامل المرافقة فكان تأخر النطق ٧١,٣% الاكثر شيوعا يليه التخلف العقلي ٦١,٥% الصرع ٣٥,٢% صعوبات سمعية ٢٦,٢% الطيف التوحدي ٤,٩% وهو الأقل، وعليه فان الطيف السريري في بلدنا يختلف عن الموجود في بلاد الغرب وهناك حاجة لمزيد من الدراسات المستقبلية الاستباقية لتقييم الطيف السريري والعوامل المهيئة للشلل الدماغي في الاردن.
- الكلمات الدالة: شلل دماغي، طيف سريري، جنوب الأردن.