

# PREVALENCE OF PJK AFTER ARTHRODESIS IN PATIENTS WITH NEUROMUSCULAR SCOLIOSIS IN THE SECOND POSTOPERATIVE YEAR

PREVALÊNCIA DE PJK APÓS ARTRODESE EM PACIENTES COM ESCOLIOSE NEUROMUSCULAR NO SEGUNDO ANO DE PÓS-OPERATÓRIO

PREVALENCIA DE PJK DESPUÉS DE ARTRODESIS EN PACIENTES CON ESCOLIOSIS NEUROMUSCULAR EN EL SEGUNDO AÑO DE POSTOPERATORIO

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## ABSTRACT

**Objective:** The primary objective of this study was to analyze the prevalence of PJK in patients with neuromuscular scoliosis submitted to posterior spinal arthrodesis with instrumentation. Proximal junctional kyphosis (PJK) is a frequent phenomenon that, due to its importance, began to be studied by several authors, who laid the foundations for the radiographic definition and possible risk factors for its occurrence after long spinal arthrodesis with instrumentation. Despite the large number of PJK studies, most were related to the occurrence of adolescent idiopathic scoliosis, adult deformity and early-onset scoliosis, with few being targeted to patients with congenital and neuromuscular scoliosis. **Methods:** In this study, data from electronic medical records of patients with neuromuscular scoliosis who underwent posterior arthrodesis with instrumentation between the years 2014 and 2016 were analyzed. Information on age, gender, pathology and radiographic measurements were extracted from this sample at the 2<sup>nd</sup> and 24<sup>th</sup> postoperative months. **Results:** A total of 39 patients with neuromuscular scoliosis were analyzed. The sample was predominantly male (58.87%) and the mean age was 14.05 years. PJK occurred in 18 patients during the two years following surgery, with a prevalence of 46.15%. The incidence of PJK in the 2<sup>nd</sup> and 24<sup>th</sup> postoperative months was 23.1% and 30%, respectively. **Conclusions:** A prevalence of PJK of 46.15% was found in patients with neuromuscular scoliosis treated surgically with posterior instrumentation after two years of follow-up, as compared to previous results. **Level of Evidence III; Cross-sectional observational study.**

**Keywords:** Kyphosis; Scoliosis; Spine.

## RESUMO

**Objetivo:** O objetivo primário deste estudo foi analisar a prevalência de PJK nos pacientes com escoliose neuromuscular, submetidos à artrodeose da coluna vertebral por via posterior com instrumentação. A cifose juncional proximal (PJK) é um fenômeno frequente que, devido à sua importância, passou a ser estudado por diversos autores, os quais lançaram as bases da definição radiográfica e possíveis fatores de risco de ocorrência depois de artrodeose longa da coluna vertebral com instrumentação. Apesar da grande quantidade de estudos sobre PJK, a maioria foi relacionada quanto à ocorrência na escoliose idiopática do adolescente, na deformidade do adulto e na escoliose de início precoce, sendo poucos direcionados a pacientes com escoliose congênita e neuromuscular. **Métodos:** Neste estudo foram analisados dados de prontuários eletrônicos de pacientes com escoliose neuromuscular submetidos à artrodeose por via posterior com instrumentação, entre os anos de 2014 e 2016. Dessa amostra, foram extraídas informações pertinentes a idade, gênero, patologia e medidas radiográficas no 2º e 24º mês de pós-operatório. **Resultados:** Um total de 39 pacientes com escoliose neuromuscular foi analisado. Na amostra houve predomínio de pacientes do sexo masculino (58,87%) e a média de idade foi de 14,05 anos. A PJK ocorreu em 18 pacientes no decorrer de dois anos de pós-operatório, com prevalência de 46,15%. A incidência de PJK no 2º e 24º mês de pós-operatório foi de 23,1% e 30%, respectivamente. **Conclusões:** Foi encontrada prevalência de 46,15% de PJK em pacientes com escoliose neuromuscular tratados cirurgicamente com instrumentação por via posterior depois de dois anos de seguimento, confrontado resultados anteriores. **Nível de Evidência III; Estudo observacional analítico transversal.**

**Descritores:** Cifose; Escoliose; Coluna Vertebral.

## RESUMEN

**Objetivo:** El objetivo primario de este estudio fue analizar la prevalencia de PJK en los pacientes con escoliosis neuromuscular, sometidos a artrodesis de la columna vertebral por vía posterior con instrumentación. La cifosis de unión proximal (PJK) es un fenómeno frecuente que, debido a su importancia, pasó a ser estudiado por diversos autores, los cuales lanzaron las bases de la definición radiográfica y posibles factores de riesgo de ocurrencia después de artrodesis larga de la columna vertebral con instrumentación. A pesar de la gran cantidad de estudios sobre PJK, la mayoría fue relacionada cuanto a la ocurrencia en la escoliosis idiopática del adolescente, en la deformidad del adulto y en la escoliosis de inicio precoz, siendo pocos direccionados a pacientes con escoliosis congénita y neuromuscular. **Métodos:**

Study conducted at the Hospital AACD, São Paulo, SP, Brazil.

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En este estudio se analizaron datos de prontuarios electrónicos de pacientes con escoliosis neuromuscular sometidos a artrodesis por vía posterior con instrumentación, entre los años 2014 y 2016. De esa muestra se extrajeron informaciones pertinentes a edad, género, patología y medidas radiográficas en el 2º y 24º mes de postoperatorio. Resultados: Fue analizado un total de 39 pacientes con escoliosis neuromuscular. En la muestra hubo predominio de pacientes del sexo masculino (58,87%) y el promedio de edad fue de 14,05 años. La PJK ocurrió en 18 pacientes en el transcurso de 2 años de postoperatorio, con prevalencia de 46,15%. La incidencia de PJK en el 2º y 24º mes de postoperatorio fue de 23,1% y 30%, respectivamente. Conclusiones: Se encontró prevalencia de 46,15% de PJK en pacientes con escoliosis neuromuscular tratados quirúrgicamente con instrumentación por vía posterior después de 2 años de seguimiento, confrontando resultados anteriores. **Nivel de evidencia III; Estudio observacional analítico transversal.**

**Descriptores:** Cifosis; Escoliosis; Columna Vertebral.

## INTRODUCTION

Proximal junctional kyphosis (PJK), which consists of a non-physiological abnormality in the sagittal plane that occurs following surgery in the segment proximal to the arthrodesis, is one of the postoperative complications that confronts the spine surgeon. In 1989, Bernhardt and Bridwell<sup>1</sup> defined the segmental angulation, as measured by the Cobb method, of each level of the spine from T1T2 to L5S1. Then in 1999, Lee et al.<sup>2</sup> defined abnormal PJK as kyphosis, measured from T2 to the proximal instrumented level,  $\geq 5^\circ$  above the sum of the normal values of each segment as previously described by Bernhardt and Bridwell.<sup>1</sup> In 2005 Glattes et al.<sup>3</sup> modified the definition of PJK to a sagittal proximal junctional Cobb angle measured between the lower vertebral endplate of the uppermost instrumented vertebra (UIV) and the upper vertebral endplate of the 2 suprajacent vertebrae that is  $\geq 10^\circ$  and at least  $10^\circ$  greater than the preoperative measurement of the segment. In 2010 Helgeson et al.<sup>4</sup> proposed another method to define PJK using the upper vertebral endplate of the adjacent vertebra and the lower vertebral endplate of the UIV as a basis. They suggested abnormality to be an angle more than two standard deviations greater than the normal segment, which in that case was  $15^\circ$ , and they redefined PJK as any increase in postoperative kyphosis  $\geq 15^\circ$ .

Currently there is no consensus around the definition of PJK, but that given by Glattes et al.<sup>3</sup> is the most commonly used definition in the literature.

The main objective of this study was to analyze the prevalence of PJK in patients with neuromuscular scoliosis who underwent instrumented posterior spinal arthrodesis.

## METHODS

In this retrospective, observational, epidemiological study, we analyzed 80 patients with neuromuscular scoliosis who had undergone spinal arthrodesis with instrumentation at a tertiary hospital between January 1, 2014 and December 31, 2016. Patient information related to the pathology, age, sex and surgical date, as well as panoramic spinal radiographs in a standing or sitting position taken preoperatively and in the 2nd and 24th months following surgery were collected from the database.

For the radiographic evaluation and PJK diagnosis we used the method proposed by Glattes et al.,<sup>3</sup> in which the sagittal proximal junctional Cobb angle (PJA), measured between the lower vertebral endplate of the uppermost instrumented vertebra (UIV) and the upper endplate of the two suprajacent vertebrae, measures  $\geq 10^\circ$  and at least  $10^\circ$  greater than the preoperative measurement of the segment. (Figure 1) The data were obtained from the electronic medical records archive. As the inclusion criterion, only patients diagnosed with neuromuscular scoliosis who were submitted consecutively to long posterior spinal arthrodesis with instrumentation between 01/01/2014 and 12/31/2016 who underwent adequate radiographical follow-up in the 2nd and 24th months were selected for this study. Patients who underwent posterior spinal arthrodesis without a diagnosis of neuromuscular scoliosis, as well as those previously submitted to spinal arthrodesis were excluded from the study.

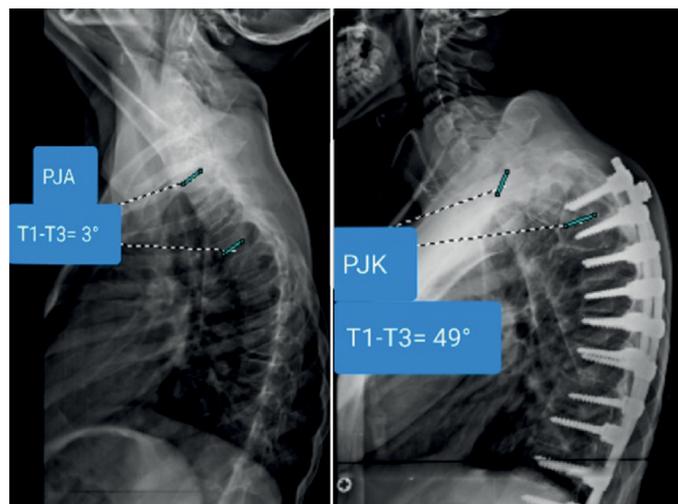
Using the criteria mentioned above, we recorded which patients had proximal junctional kyphosis  $\geq 10^\circ$  and progression of at least  $10^\circ$  greater than the preoperative measurement in the 2nd and 24th months following surgery.

For the descriptive evaluations and analytical tests, SPSS version 20.0.0 software was used. The descriptive analyses included frequency, mean and median measurements. This study was submitted to the Institutional Review Board, which waived the informed consent form (ICF) requirement, and was approved as CAEE number 08114019.8.0000.0085.

## RESULTS

A total of 39 individuals met the study inclusion criteria. Neuromuscular scolioses secondary to various pathologies were included in the study, as shown in Table 1. There was a predominance of male patients (58.87%) (Table 2) and the minimum and maximum ages were 8 and 24 years, with a mean age of 14.05 years.

The minimum and maximum preoperative PJA values measured were  $-14^\circ$  and  $17^\circ$  (one of the cases of PJA measured was negative because the patient was lordotic) with a mean of  $6.28^\circ$ . (Table 3) After 2 years of postoperative follow-up, PJK was observed in 18 of the 39 patients (prevalence of 46.15%) (Table 4). The incidence of PJK was lower at 2 months than at 2 years following surgery, with nine individuals in each group for incidences of 23.1% and 30%, respectively. (Tables 5 and 6)



**Figure 1.** Preoperative proximal junctional angle on the left and progression to proximal junctional kyphosis on the right.

**Table 1.** Frequency by diagnosis.

Diagnosis	Frequency	Percentage (%)	Cumulative	Cumulative percentage (%)
Congenital Muscular Dystrophy	2	5.128205	2	5.128205
Charcot Disease	1	2.564103	3	7.692308
Myelomeningocele	3	7.692308	6	15.384615
Central Core Myopathy	1	2.564103	7	17.948718
Mitochondrial Myopathy	1	2.564103	8	20.512821
Cerebral Palsy	30	76.923077	38	97.435897
Leigh Syndrome	1	2.564103	39	100
Total = 39				

**Table 2.** Frequency by sex.

Sex	Frequency	Percentage (%)	Cumulative	Cumulative percentage (%)
female	16	41.025641	16	41.025641
male	23	58.974359	39	100
Total = 39				

**Table 3.** Frequency of the preoperative PJA.

Value	Frequency	Percentage (%)	Cumulative	Cumulative percentage (%)
-14	1	2.564103	1	2.564103
2	4	10.25641	5	12.820513
3	2	5.128205	7	17.948718
4	4	10.25641	11	28.205128
5	6	15.384615	17	43.589744
6	3	7.692308	20	51.282051
7	2	5.128205	22	56.410256
8	7	17.948718	29	74.358974
9	4	10.25641	33	84.615385
10	1	2.564103	34	87.179487
12	4	10.25641	38	97.435897
17	1	2.564103	39	100
Total = 39				

**Table 4.** Prevalence of PJK two years following surgery

PJK	Frequency	Percentage (%)	Cumulative	Cumulative percentage (%)
N	21	53.846154	21	53.846154
Y	18	46.153846	39	100
Total = 39				

**Table 5.** Incidence of PJK two months after surgery.

PJK	Frequency	Percentage (%)	Cumulative	Cumulative percentage (%)
N	30	76.923076	30	76.9
Y	9	23.076924	39	100
Total 39				

**Table 6.** Incidence of PJK two years after surgery.

PJK	Frequency	Percentage (%)	Cumulative	Cumulative percentage (%)
N	21	70	21	30
Y	9	30	30	100
Total 30				

The mean PJK angle at two months following surgery was 28.7°, with a minimum of 19° and a maximum of 37°, and at two years following surgery was 26.3°, with a minimum of 11° and a maximum of 54°.

## DISCUSSION

PJK has been evaluated extensively over the years, both to define the diagnosis and to establish epidemiological values and risk factors for its appearance. However, most studies are conducted in patients with spinal deformities of various etiologies (AIS – adolescent idiopathic scoliosis, ASD – adult spinal deformity, Scheuermann's disease), which are not neuromuscular, or with early onset scoliosis (EOS), with very few of them conducted exclusively in patients with scoliosis of neuromuscular etiology.

Over the course of time, several studies have demonstrated a variance in the prevalence and incidence of PJK. In 1999, Lee et al.<sup>2</sup> reported a prevalence of PJK of 46% of the 69 patients treated surgically for AIS. Later studies clarified both risk factors and the prevalence and incidence of PJK in AIS, ASD and EOS.<sup>3-5</sup> Kim et al.<sup>6</sup> showed that the prevalence of PJK after an average follow-up of 7.3 years was 26%, with a diagnosis in 82% up to the second postoperative year and without significant progression after two years following surgery in patients with AIS, which was later corroborated by Kim et al.<sup>5</sup> in a new study with 410 cases and a prevalence of 27%. In the same way, similar results were observed by Glattes et al.<sup>3</sup> and in other studies of ASD.<sup>7,8</sup>

In EOS, a prevalence of around 26%, like that of PJK, was also confirmed in several other studies, both of patients treated with arthrodesis and with treatments using growth-guided devices.<sup>8-11</sup> Chunda et al.<sup>12</sup> reported a PJK prevalence of 11%. But it was a meta-analysis of children and adolescents with spinal deformity of any etiology who had undergone surgical treatment. The emergence of PJK occurs in the first months<sup>13-15</sup> and after the 2nd year following surgery there is little progression, with 66% of cases being diagnosed by the 3rd postoperative month and 80% within a year and a half following surgery.<sup>15</sup>

As mentioned previously, few studies have been conducted exclusively in patients with neuromuscular scoliosis and these show divergent prevalence and incidence rates. In 2003 and 2012, Sink et al.<sup>16</sup> and Lonstein et al.<sup>17</sup> published studies of patients with neuromuscular scoliosis, mainly cerebral palsy, reporting PJK in four of 41 and two of 93 patients studied, respectively.

This study showed a prevalence of PJK of 46.15% in patients with neuromuscular scoliosis who underwent corrective surgery with posterior spinal instrumentation, like the study conducted by Lee et al.,<sup>2</sup> but in patients with AIS. In relation to more recent studies,<sup>3,5-11</sup> which obtained a prevalence of around 26% in patients with AIS, ASD and EOS, the prevalence observed was higher. The incidence of PJK two months following surgery (23.1%) was less than in the second postoperative year (30%), there being nine individuals in each group, but in studies by Chen et al.<sup>13</sup> and Lau et al.<sup>15</sup> PJK was diagnosed in 66 to 90% of cases in the first three months following treatment, which differed from our PJK diagnosis results of 50% in the first two months following surgery. In the few studies conducted in patients with neuromuscular scoliosis, the prevalence of PJK was less than that found in our study,<sup>12,16,17</sup> although in these studies the patients analyzed were submitted to less rigid spinal instrumentation and the radiographic standards for a diagnosis of PJK were different than those used in our study.

## CONCLUSION

The prevalence of PJK in patients with neuromuscular scoliosis was 46.15% two years after surgery. There was a difference in incidence in the 2nd and 24th postoperative months of 23.1% and 30%, respectively, with the incidence of scoliosis in the 2nd month following surgery consistent with the other studies, although they were conducted with patients with AIS, ASD and EOS.

After conducting this study, we see the need for more research focused in the emergence of PJK in patients with neuromuscular scoliosis given the lack of recent studies with the new instrumentation materials in long posterior spinal arthrodesis and the conflicting results in the literature in relation to the prevalence of PJK.

All authors declare no potential conflict of interest related to this article.

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