Kyphectomy in Patients with Myelomeningocele. Is there a Better Age for Surgery Correction?

Cifectomia em Pacientes com Mielomeningocele. Existe Uma Idade Melhor para a Correção da Cirurgia?

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ABSTRACT

Introduction: myelomeningocele can be associated with spine curve deformities, including kyphosis. These deformities can be progressive and impair patients’ quality of life. Surgery for correction is often necessary. However, there is no consensus on when the surgical procedure should be performed. Thus, the objective of this analysis is to compare kyphectomy in different age groups. Methods: systematic review of literature, performed by PRISMA protocol. The PICOT strategy was used, where P - patients with myelomeningocele associated with kyphosis, I - for neonatal kyphectomy, C - patients who did not undergo neonatal kyphectomy but who underwent it at another age, and T - follow-up period. Results: overall, 21 articles reporting 142 patients were analyzed. The patients were subdivided into three groups according to the age of kyphosis surgery: neonatal, up to ten years old, and over to ten years old. The mean follow-up was 3.8 years (2.3-5.9), and the mean kyphosis corrected angle was 84º (SD = 34º). All subjects studied showed statistical significance in the comparison between pre-and-post-operative kyphosis angles, reinforcing the effectiveness of surgical therapy for this deformity. In our results, all groups presented with an improvement in the quality of life (QoL), nevertheless the neonates had a higher rate of QoL. In our analysis, there was no difference between groups during the safety assessment. No neonatal deaths were reported. In comparison, the groups ‘up to 10 years’ and ‘over to 10 years’ had a mortality rate of 1.4% and 0.7%, respectively. Conclusion: the present study shows that kyphectomy is the surgical procedure of choice for treating kyphosis in patients with myelomeningocele and it is safe and effective in all age groups. In addition, the procedure performed on neonates seems to improve the QoL and if necessary, a late second surgery will be more straightforward.

Keywords: Myelomeningocele; Neonate; Kyphosis correction angle; Quality of life; Kyphosis

RESUMO

Introdução: a mielomeningocele pode estar associada a deformidades nas curvas da coluna, incluindo a cifose. Essas deformidades podem ser progressivas e prejudicar a qualidade de vida dos pacientes. Muitas vezes, a cirurgia para correção é necessária. No entanto, não há consenso sobre quando o procedimento cirúrgico deve ser realizado. Assim, o objetivo desta análise é comparar a cifectomia em diferentes faixas etárias. Métodos: revisão sistemática da literatura, realizada pelo protocolo PRISMA. Foi utilizada a estratégia PICOT, onde P - pacientes com mielomeningocele associada à cifose, I - para cifectomia neonatal, C - pacientes que não realizaram cifectomia neonatal, mas que a realizaram em outra idade, e T - período de acompanhamento. Resultados: ao todo, 21 artigos relatando 142 pacientes foram analisados. Os pacientes foram subdivididos em três grupos de acordo com a idade da cirurgia de cifose: neonatal, até dez anos e acima de dez anos. O seguimento médio foi de 3,8 anos (2,3-5,9), e o ângulo médio de correção da cifose foi de 84º (DP = 34º). Todas as populações estudadas apresentaram significância estatística na comparação entre os ângulos de cifose pré e pós-operatórios, reforçando a eficácia da terapia cirúrgica para essa deformidade. Em nossos resultados, todos os grupos apresentaram melhora na qualidade de vida (QV), porém os neonatos apresentaram maior índice de QV. Em nossa análise, não houve diferença entre os grupos durante a avaliação de segurança. Nenhuma morte neonatal foi relatada. Em comparação, os grupos de até 10 anos e acima de 10 anos apresentaram taxa de mortalidade de 1,4% e 0,7%, respectivamente. Conclusão: o presente estudo mostra que a cifectomia é o procedimento cirúrgico de escolha para o tratamento da cifose em pacientes com mielomeningocele, sendo seguro e eficaz em todas as faixas etárias. Além disso, o procedimento realizado em recém-nascidos parece melhorar a qualidade de vida e, se necessário, uma segunda cirurgia tardia será mais direta.

Palavras-chave: Mielomeningocele; Neonato; Ângulo de correção da cifose; Qualidade de vida; Cifose
**INTRODUCTION**

Myelomeningocele (MMC) is commonly associated with spine deformities and affects approximately 10% to 15% of neonates with this condition\(^1,2\). Of them, kyphosis represents the main one, and it can evolve with growth disorders, skin ulcers, respiratory disturbances, and an overall loss of QoL\(^3,4\).

The first reported neonatal kyphectomy was performed in 1968 by Sharrard\(^5\). Since then, many surgical techniques have been described, which forwarded early correction of kyphosis in patients with MMC\(^2,6\). Despite an initial restriction to perform this procedure during the early days of life, due to blood loss concerns, the number of procedures in neonates has increased. The advances in hemostatic techniques have made this procedure safer. Nonetheless, there is still no consensus on the best age to correct kyphosis.

Thus, the objective of this study is to assess whether there is a difference in the kyphosis correction angle between neonates, and children younger and older than 10 years old. The study also assessed the procedure’s safety in the different age groups.

**METHODS**

This study is a systematic review of literature, performed by PRISMA protocol. The PICOT strategy was used, where P - patients with myelomeningocele associated with kyphosis, I - for neonatal kyphectomy, C - patients who did not undergo neonatal kyphectomy but who underwent it at another age, and T - follow-up period. Only patients who underwent kyphectomy were included in this analysis. Studies using secondary data sources were excluded.

We searched the Medline (via PUBMED), Web of Science, and SciELO databases for clinical trials randomized or not, case reports, and case series between January 1979 and October 2020, on the topic of kyphectomy in patients with myelomeningocele published in English, Portuguese, or Spanish. The keywords used for the search were “myelomeningocele”, “kyphectomy” and “neonatal kyphosis”.

The search identified 57 articles, of these 21 fulfilled the inclusion criteria and were analyzed (Figure 1).

![Research and literature screening flow diagram](image)

*Figure 1. Research and literature screening flow diagram.*
The variables collected were: pre-operative kyphosis angle (pre-KA), post-operative kyphosis angle (post-KA), skin ulcers, lower limb sensitivity, ability to be seated, level of spine deformity, death, and follow-up period.

The correction angle was defined as the difference between pre-and post-operative kyphosis angles.

QoL was assessed by the improvement to lower limb sensitivity, acquired the ability to remain seated, and the absence of skin ulcers. According to the patient’s response, he would be categorized into three groups: improved, unchanged, and worsened.

**Statistical analysis**

To analyze categorical variables, absolute and relative frequencies were performed. For continuous variables, data were presented according to normality, with mean and standard deviation for variables with normal distribution and median and their quartiles in non-normal data.

To analyze the difference between values of preoperative and postoperative kyphosis angle the paired t-test was performed between the same groups. To compares the correction angle of kyphosis between the three groups, One Way-ANOVA was used and the pairwise comparison was necessary to describe which group had a significant statistical difference.

The Kruskal Wallis test was performed to check whether there was a statistical difference between the groups in terms of kyphosis levels. The p values under 0.05 were considered as statistically significant. All analyses were performed with the Statistical Package for Social Sciences (IBM SPSS Statistics for Windows, version 20.0; IBM, Armonk, New York) software.

**RESULTS**

Overall, 21 articles reporting 142 patients were analyzed. The median follow-up was 3.8 years (2.3-5.9) and the mean corrected angle was 84º (SD = 34º).

For descriptive purposes, the patients were categorized in three groups: neonatal - 12% (n= 17); up to 10 years old – 61.3% (n=87); and over 10 years old – 26.8% (n=38). The comparison between the age groups was described in Table 1.

The mean age of the surgical procedure was 5.6 days (SD 3.33) in the neonatal group, and in ‘over to 10 years’ was 13.4 years (SD 2). The median in ‘up to 10 years’ was 7 years (IQR 6-7.1).

The follow-up was statistically significant in the Kruskal Wallis test (p = 0.02). The pairwise comparison showed that the neonatal group had a higher period of follow-up compared to ‘over 10 years’ group (p = 0.037).

### Table 1. Comparison between age groups.

<table>
<thead>
<tr>
<th></th>
<th>Neonatal</th>
<th>Up to 10 years</th>
<th>Over to 10 years</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Follow-up (years)</strong>*</td>
<td>4.9 (3.55 – 5.95)</td>
<td>3.95 (2.55 – 6.95)</td>
<td>3.1 (1.5 – 4.7)</td>
<td>0.02</td>
</tr>
<tr>
<td><strong>Level</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Thoracic</td>
<td>5.9% (1)</td>
<td>19.5% (17)</td>
<td>26.3% (10)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Thoracolumbar</td>
<td>0%</td>
<td>23% (20)</td>
<td>36.8% (14)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Lumbar</td>
<td>94.1% (16)</td>
<td>57.5% (50)</td>
<td>36.8% (14)</td>
<td></td>
</tr>
<tr>
<td><strong>Skin ulcer</strong></td>
<td>0%</td>
<td>12.6% (11)</td>
<td>7.9% (3)</td>
<td></td>
</tr>
<tr>
<td><strong>Quality of life</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Improved</td>
<td>100% (17)</td>
<td>97.7% (85)</td>
<td>89.5% (24)</td>
<td></td>
</tr>
<tr>
<td>Unchanged</td>
<td>-</td>
<td>-</td>
<td>2.6% (1)</td>
<td></td>
</tr>
<tr>
<td>Worsened</td>
<td>-</td>
<td>2.3% (2)</td>
<td>7.9% (3)</td>
<td></td>
</tr>
<tr>
<td>Mortality</td>
<td>0%</td>
<td>1.4% (2)</td>
<td>0.7% (1)</td>
<td></td>
</tr>
</tbody>
</table>

*Median (interquartile range); all other variables: % (n).
The lumbar kyphosis prevailed among the three groups, representing 94.1% of the level in the neonatal group. The Kruskal Wallis test demonstrated that there was a difference between the thoracic and lumbar levels of kyphosis in the groups (p < 0.001).

Regarding the degrees of correction of kyphosis (Table 2), there was an average reduction of 37.6 (CI 95% 28.7 – 46.6) and the paired sample t-test demonstrated there was a statistical difference between pre-KA and post-KA in the neonatal group (p < 0.01). The same was observed in the ‘up to 10 years’ group, with an average reduction of 92.9 (CI 95% 86.5 – 99.3), significant statistically (p < 0.01), and ‘over 10 years’ with a reduction of 84.6 (CI 95% 73.8 – 95.3, p < 0.01).

For analysis between age groups, it was used the One-way ANOVA, and a different corrected angle of kyphectomy was demonstrated in the neonatal group when compared with ‘up to 10 years’ and ‘over 10 years’ (p < 0.05, p < 0.05, respectively). However, there was no difference between ‘up to 10 years’ and ‘over 10 years’ (p = 0.32).

Results per study were presented in Table 3.

**DISCUSSION**

Neonatal kyphectomy in patients with myelomeningocele was first proposed by Sharrard, in 1968, with the procedure performed successfully and without complications. Furthermore, in the same study, correction of the kyphosis angle was maintained, during the follow-up, in approximately 67% of the patients. In cases of recurrence, it was possible to use splints over the residual kyphosis in older children, which contributed to the decrease in the probability of kyphosis recurrence and need for reoperation.

Before Sharrard’s study, conservative treatment was attempted to stabilize the kyphosis angle. These methods aim to reduce the pressure on the hump, being adapted to wheelchairs, seat cushions, beanbags, and braces. These strategies are considered ineffective, as the degree of kyphosis tends to progress quickly, causing the patient to incline the trunk, more and more, and use the upper limbs as crutches. Another major complication studied in clinical outcomes is hump ulcers, which are associated with long periods in the supine position. They are associated with both a gateway to opportunistic infections and increased intra-abdominal pressure, which leads to decreased appetite and loss of pulmonary reserve.

So far, after extensive research in the literature, only 34 reported cases of neonatal kyphectomy were found, 17 of which were included in this research because they met the inclusion and exclusion criteria. This low number available is explained by the fact that neonatal kyphectomy was discouraged for decades due to the high mortality rate found in previous studies. However, this high mortality rate was described more than 4 decades ago, when the technology did not support better synthesis and hemostasis methods, such as mono- and bipolar electrocautery. Additionally, Sharrard and Drennan reported that of the 3 deaths in his initial series, two were unrelated to the surgical procedure.

It was only after the 1970s that neonatal kyphectomy began to be demystified. Eckstein and Vora, in 1972, considered that the surgical procedure in neonates is simpler when compared to older children. His series evaluated 16 patients, 1 of which was a neonate.

One of the main variables that were associated with the worst outcome in the neonatal group was operative bleeding. This variable can be controlled in neonatal kyphectomies with the use of hypotensive anesthesia and with electrocautery.

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**Table 2. Comparison between age groups of kyphotic angle.**

<table>
<thead>
<tr>
<th>Kyphotic angle</th>
<th>Neonatal</th>
<th>Up to 10 years</th>
<th>Over to 10 years</th>
</tr>
</thead>
<tbody>
<tr>
<td>Preoperative Kyphosis angle</td>
<td>71º (16)*</td>
<td>120º (100-150)*</td>
<td>126.9º (34.2)*</td>
</tr>
<tr>
<td>Postoperative Kyphosis angle</td>
<td>33.4º (17)*</td>
<td>25.5º (15-41.2)*</td>
<td>43º (24-57.5)*</td>
</tr>
</tbody>
</table>

*Mean (SD). *Median (interquartile range).
Table 3. Details of the included 142 patients of kyphosis and myelomeningocele.

<table>
<thead>
<tr>
<th>Author</th>
<th>Mean Age (Years)</th>
<th>Kyphotic Angle</th>
<th>Quality of Life</th>
<th>Follow-up</th>
<th>Mean Age (Years)</th>
<th>Kyphotic Angle</th>
<th>Quality of Life</th>
</tr>
</thead>
<tbody>
<tr>
<td>Böhm and ElSaghir, 2000</td>
<td>10.0</td>
<td>92.0</td>
<td>23.00</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>NR</td>
</tr>
<tr>
<td>Comstock et al., 2010</td>
<td>7.5</td>
<td>123.2</td>
<td>40.43</td>
<td>21</td>
<td>1</td>
<td>0</td>
<td>NR</td>
</tr>
<tr>
<td>Gepp et al., 2013</td>
<td>10.0</td>
<td>116.3</td>
<td>51.87</td>
<td>8</td>
<td>0</td>
<td>0</td>
<td>NR</td>
</tr>
<tr>
<td>Ganjeifar et al., 2016</td>
<td>15.0</td>
<td>137.0</td>
<td>30.00</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>NR</td>
</tr>
<tr>
<td>Garg et al., 2011</td>
<td>12.0</td>
<td>147.1</td>
<td>50.89</td>
<td>17</td>
<td>0</td>
<td>0</td>
<td>NR</td>
</tr>
<tr>
<td>Kiepe et al., 2019</td>
<td>5.0</td>
<td>180.0</td>
<td>NR</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>NR</td>
</tr>
<tr>
<td>Ko et al., 2007</td>
<td>10.8</td>
<td>124.6</td>
<td>37.67</td>
<td>8</td>
<td>1</td>
<td>0</td>
<td>NR</td>
</tr>
<tr>
<td>Kocaoğlu et al., 2008</td>
<td>7.1</td>
<td>104.0</td>
<td>15.29</td>
<td>7</td>
<td>0</td>
<td>0</td>
<td>NR</td>
</tr>
<tr>
<td>Laing et al., 2006</td>
<td>3.0</td>
<td>110.0</td>
<td>20.00</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>NR</td>
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<tr>
<td>Miyamoto et al., 2003</td>
<td>9.0</td>
<td>113.0</td>
<td>10.00</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>NR</td>
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<tr>
<td>Alshaalan et al., 2019</td>
<td>3.0</td>
<td>155.0</td>
<td>85.00</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>NR</td>
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<tr>
<td>Odent et al., 2004</td>
<td>9.7</td>
<td>111.1</td>
<td>16.11</td>
<td>8</td>
<td>1</td>
<td>0</td>
<td>NR</td>
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<tr>
<td>Ryabykh et al., 2018</td>
<td>4.6</td>
<td>94.6</td>
<td>27.55</td>
<td>11</td>
<td>0</td>
<td>0</td>
<td>NR</td>
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<tr>
<td>Smith et al., 2016</td>
<td>2.0</td>
<td>120.0</td>
<td>57.00</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>NR</td>
</tr>
<tr>
<td>Thomsen et al., 2000</td>
<td>8.0</td>
<td>152.4</td>
<td>44.22</td>
<td>9</td>
<td>0</td>
<td>0</td>
<td>NR</td>
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<tr>
<td>Crawford et al., 2003</td>
<td>Neonates</td>
<td>67.0</td>
<td>27.56</td>
<td>9</td>
<td>0</td>
<td>0</td>
<td>NR</td>
</tr>
<tr>
<td>Dunn and Bomela, 2016</td>
<td>9.8</td>
<td>143.4</td>
<td>18.57</td>
<td>7</td>
<td>0</td>
<td>0</td>
<td>NR</td>
</tr>
<tr>
<td>Kaplan et al., 2015</td>
<td>5.6</td>
<td>114.3</td>
<td>28.17</td>
<td>4</td>
<td>2</td>
<td>0</td>
<td>NR</td>
</tr>
<tr>
<td>Özdemir et al., 2019</td>
<td>Neonates</td>
<td>75.6</td>
<td>40.00</td>
<td>8</td>
<td>0</td>
<td>0</td>
<td>NR</td>
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<tr>
<td>Samagh et al., 2011</td>
<td>9.0</td>
<td>115.6</td>
<td>13.00</td>
<td>11</td>
<td>0</td>
<td>0</td>
<td>NR</td>
</tr>
<tr>
<td>Wui et al., 2019</td>
<td>5.0</td>
<td>179.0</td>
<td>35.00</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>NR</td>
</tr>
</tbody>
</table>

*In years; NR: Not Reported.*
In our study, the selected patients were followed for a long period. The neonatal population had the highest mean follow-up, followed by the 'up to 10 years' and 'over to 10 years' group, respectively (4.9, 3.95, 3.1, p = 0.02).

Furthermore, all populations studied showed statistical significance in the comparison between pre- and post-kyphectomy kyphosis angles, reinforcing the effectiveness of surgical therapy for this deformity. This reduction was also demonstrated in the Crawford study in neonates, in which he presented a preoperative mean of 67º and an initial correction mean of 77º. All studies included in this analysis in which non-neonatal patients were included also demonstrated a significant reduction in the degree of kyphosis and an improvement in the QoL of most of their patients,2,3,6,7,11-14,23,24.

Another factor compared in our research was the degree of correction between groups, in which the neonates had a lower average than the other children studied. Furthermore, a statistically significant difference was demonstrated between the neonatal group and the others (p < 0.05) in the angle of correction. However, no difference was demonstrated between the groups up to 10 years and above 10 years. This can be explained by the lower averages of the degrees of kyphosis presented in the preoperative period in the neonatal, 'up to 10 years' and 'over 10 years' groups, being 71º, 120º, and 126.9º, respectively.

Skin lesions were evaluated as they constitute an important predictor of QoL. In our results, no postoperative skin lesions were reported in the neonatal group until the end of the follow-up period. In the group 'up to 10 years old', the prevalence was 12.6% and in those 'over 10 years old', it was 7.9%.

Özdemir et al.2 in a series of 8 cases of kyphectomy in neonates with myelomeningocele, reported that there was no need for blood transfusion in any of his patients during the surgical procedure. In our analysis, it was not possible to compare the blood loss during the surgical procedure between the studies, as the data were very heterogeneous or their description was absent.

In our results, the QoL assessment was considered improved in all neonates. There are no other literature reviews that address this issue, but these results were also found in case series not included in the research, such as Crawford et al.6, in which all patients who had previously undergone neonatal kyphectomy were able to remain seated comfortably and maintain the use of wheelchairs.

The surgical procedure was safe in our analysis in different age groups. No neonatal deaths were reported. In comparison, the 'up to 10 years' and 'over to 10 years' group had a higher, albeit low, mortality rate (1.4% and 0.7%, respectively).

This study has some limitations. First, as this is a systematic review of a series of cases or reports, the limitations regarding the design of observational studies must be taken into account. Second, not all studies in the neonatal population could be included due to lack of information or insufficient description of patient data, such as follow-up, age, degrees of kyphosis, mortality, and factors necessary to assess the improved quality of life. Furthermore, the number of cases of neonatal kyphectomy in patients with MMC is limited.

CONCLUSION

The present study shows that kyphectomy is the surgical procedure of choice for treating kyphosis in patients with myelomeningocele and being safe and effective in the different ages analyzed. In addition, the procedure earlier performed seems to improve the quality of life in the neonatal population.

REFERENCES


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