Displasia do desenvolvimento do quadril: série de casos no Rio Grande do Norte

Hip developmental dysplasia: case report in Rio Grande do Norte

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ABSTRACT: Developmental dysplasia of the hip (DDH) represents a spectrum of anatomical abnormalities of the hip. The objective of this manuscript is to report the atypical presentation pattern of developmental dysplasia of the hip present in a family group in Rio Grande do Norte and to emphasize the importance of early diagnosis performed through the Ortolani and Barlow maneuvers. Both orthopedic clinical tests are essential in screening for changes in the hip joint in the newborn and should preferably be performed from birth to approximately three months of age. In addition, requesting complementary ultrasound of the hip could be indicated. Pediatric patients with developmental hip dysplasia treated at the Children’s Rehabilitation Center were atypical clinical cases because they had male predominance in a family group in Rio Grande do Norte/RN, Brazil. It is concluded that this report emphasizes the need for early and adequate clinical evaluation by the physician for all newborns, regardless of gender, as well as the creation of a protocol which meets the suspicion parameters for requesting an ultrasound of the hip joint in newborns in order to diagnose and treat this condition early, thus reducing the need for surgical treatment and its complications.

KEY WORDS: Developmental Dysplasia of the Hip; Hip Dislocation, Congenital; Bone Diseases, Developmental.

RESUMO: A Displasia do Desenvolvimento do Quadril (DDQ) representa um espectro de anormalidades anatômicas do quadril. Nessa direção, o objetivo deste manuscrito é relatar o padrão de apresentação atípica da displasia do desenvolvimento do quadril presente em um grupo familiar no Rio Grande do Norte e enfatizar a importância do diagnóstico precoce, realizado por intermédio das manobras de Ortolani e Barlow. Ambos os testes clínicos ortopédicos são essenciais no rastreamento de alterações da articulação coxofemoral no recém-nascido e devem ser executados, preferencialmente, desde o nascimento até, aproximadamente, os três meses de idade. Além da solicitação complementar de exames ultrassonográficos do quadril, quando indicados. Os pacientes pediátricos com displasia do desenvolvimento do quadril atendidos no Centro de Reabilitação Infantil (CRI) foram casos clínicos atípicos, por apresentarem predominância masculina em um grupo familiar no Rio Grande do Norte/RN, Brasil. Conclui-se que este relato enfatiza a necessidade de avaliação clínica precoce e adequada do médico para todos os recém-nascidos, independentemente do sexo, como também a criação de um protocolo que atenda parâmetros de suspeição para a solicitação de ecografia da articulação coxofemoral em recém-nascidos, com o intuito de diagnosticar e tratar precoce e, assim reduzindo a necessidade de tratamento cirúrgico e suas complicações.

PALAVRAS-CHAVE: Displasia do Desenvolvimento do Quadril; Luxação Congênita do Quadril; Doenças do Desenvolvimento Ósseo.

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INTRODUCTION

According to the Brazilian Hip Society (2022), the Greek terms dys, which refers to the word bad or mal, and plasis, which means formation, originated the word Dysplasia. According to Guarniero (2010, p. 117), hip dysplasia encompasses abnormalities in the size, morphology or anatomical orientation, or in the organization of the acetabulum, femoral head or both.

Developmental Dysplasia of the Hip (DDH) is a term that describes a spectrum of anatomical abnormalities of the hip, meaning that it covers joint disorders between the acetabulum and the proximal femur. These can be subdivided into congenital or developmental after birth.

There is a dependent relationship between the femur and the acetabulum in forming the hip joint. Therefore, any interference with adequate contact between these two structures during pregnancy or childhood can lead to DDH. In fact, the hip joint is already recognizable in the 11th intrauterine week. However, the growth of the femoral head is faster than the growth of the acetabulum during pregnancy, which can lead to insufficient coverage of the femoral head.

The risk factors for DDH are: Italians and their descendants, female sex (ratio of 8 girls to 1 boy), firstborn, white race, young mother, family history, breech presentation at birth, oligohydramnios, occurrence of congenital torticollis, plagiocephaly, metatarsus varus, calcaneus valgus, and knee extension contracture. Goiano also reinforces the fact that the literature describes DDH as more common in females, especially when there are known risk factors.

The causes of DDH are multifactorial, but the most notable are ligament hyperlaxity and intrauterine malposition. It is important to note that the femur of the fetus in flexion and external rotation can be forced out of the acetabulum during the last months of pregnancy in the breech position, predisposing the child to be born with a dislocated, unstable or subluxed hip, as shown in Figure 1.

The ideal age range for both diagnosis and treatment is between birth (newborn) and approximately three months of age, as the prognosis for DDH is better.

The objective of this study is to report the atypical presentation pattern of DDH present in a family group in Rio Grande do Norte, Brazil, and emphasize the importance of early diagnosis.

CASE SERIES

The family consists three siblings born by normal birth, at term, without complications. They presented adequate NPMD (Neuropsychomotor Development) and were treated at the Children’s Rehabilitation Center (Centro de Reabilitação Infantil - CRI) in the city of Natal, located in the state of Rio Grande do Norte, Northeast Brazil. The first child, LOLQ, five years old, male, presented gait with an increased base of support, hyperlordosis and difficulty abducting the hips. After radiography, the patient was diagnosed with DDH (Figure 2a and 2b).

He underwent open reduction, femoral shortening and Salter-type pelvic osteotomy on the right, remaining in a pelvic-foot cast for six weeks. After six months, he underwent the same procedure on his left hip (Figures 3a, 3b, 4a, 4b).
Figure 2a and 2b - LOLQ at five years old and bilateral developmental dysplasia of the hip. Preoperative radiographs

Figure 3a and 3b - First stage of surgical correction

Figure 4a and 4b - Second stage of surgical correction. Open reduction by Salters-type pelvic osteotomy (complete transiliac osteotomy), showing good acetabular coverage in both hips. On the right, the head of the femur presents changes suggestive of sequelae of avascular necrosis, a common complication in this procedure.

The second child, BLQ, three years old, male. He was examined at one month of age, showing bilateral hip dysplasia (Figure 5a and 5b), undergoing treatment using a Pavlik harness (Figure 6a and 6b), showing good acetabular remodeling.

Figure 5a and 5b - BLQ, 3 years old, presenting bilateral DDH

Figure 6a and 6b - Pavlik harness treatment for bilateral hip dysplasia in a three-year-old male.
The third child, MQL, one month old, female, physical examination was unremarkable. USG of the hips (Figure 7) was performed within normal limits (lateral bone margin of the acetabulum angled, type 1a hips according to Graf’s method, right and left bone roof angle, 70.2 and 70.8 degrees, respectively, and right and left cartilaginous roof angle, 44 and 46.1 degrees, respectively).

**Figure 6a and 6b** - BLQ, 3 years old, after successful use of the Pavlik harness

**Figure 7** - Ultrasound of children’s hips. Right and left hip with angulated lateral bone margin of the acetabulum. Type 1a hips according to the Graf method. Bone roof angle (alpha): 70.2 degrees right and 70.8 degrees left. Cartilaginous roof angle (beta): 44 degrees right and 46.1 degrees left
### DISCUSSION

The DDH incidence in relation to dislocated hips is approximately one in every 1,000 newborns, while the incidence for subluxated (unstable) hips is approximately 10 in 1,000. It is important to highlight that geographic location influences the number of cases.

An incidence of approximately 5 cases per 1,000 newborns is expected in terms of Ortolani sign positivity in Brazil. The profile with the highest incidence in relation to Brazilian epidemiology is: female, white and with right-sided involvement (47.8%). However, the highest incidence in unilateral situations in the United States (60%) occurs in the left hip, while the least affected, 20%, is the right hip. Bilaterality occurs less frequently in 20% of cases.

There are several etiopathogenesis for DDH: intrauterine position, hormonal factors, position after birth and genetic factors. The present case report is strongly associated with etiopathogenesis due to genetic factors and a DDH occurrence pattern can be observed, since only male children manifested it, which goes against the pattern described by Hebert et al. and Barros Filho et al.

DDH can be classified according to hip instability (typical classification) into four types: dislocation, subluxation, “luxable” hip, “subluxable” hip. Furthermore, there is also the classification of teratological dislocation, when there is hip dislocation associated with other morphological changes, making reduction more difficult. This group includes only dislocated hips, which are associated with arthrogryposis, Larsen syndrome, variants of proximal femoral deficiency, Ehlers-Danlos syndrome and other syndromes. Neuromuscular instability appears as a separate classification, which in this group are hip instabilities and dislocations associated with neuromuscular diseases, such as myelomeningocele, cerebral palsy and sacral agenesis.

There is the Tönnis classification for idiopathic cases, which has limitations in younger children due to the absence of an ossification nucleus. Therefore, the International Hip Dysplasia Institute (IHDI) developed a classification that can be applied to children of all ages (Figure 8), as it is based on ultrasound findings (USG) which enables visualizing the cartilaginous femoral head, as the femoral head is not visible in newborns and young infants on radiographs.

The main limitation of the IHDI classification is the two-dimensional assessment obtained from an anteroposterior view of the pelvis, as assessment in a single plane is not sufficient to reproduce a more reliable image of the acetabulum. Some radiographic parameters, such as the Wiberg center-edge angle and the Hilgenreiner acetabular angle, are used to quantify subtle differences in acetabular dysplasia. Wiberg’s center-edge angle is used in older children to assess the degree of the femoral head lateral coverage by the acetabulum in the frontal plane. The Hilgenreiner acetabular index quantifies acetabulum development by measuring the inclination of the bony acetabulum in the frontal plane.

The diagnosis of DDH in newborns and babies is clinical and carried out with the Ortolani (hip reduction maneuver) and Barlow maneuver (hip dislocation maneuver). Ortolani’s sign: the maneuver is performed with the child in a horizontal supine position, calm and motionless, with the hips and knees in a 90° flexion position, with the thighs in adduction and with slight medial rotation. One hip should be examined at a time. Rest the thumb on the inner surface of the thigh, the palm of the hand on the knee and leg, and the index and middle fingers on the greater trochanter. Abduction and slight external rotation of the hip must be performed in the initial slight internal rotation position. The test is positive when a “clunk” of hip reduction is heard, which is substantially different from the “click” generally heard. The “clunk” is the sensation of the femoral head sliding, as it is reduced with a dull click. Other clicking sounds can be heard, usually in the knees, or even in the hip, but this does not mean a positive test. The test is negative in irreducible and rigid hips (especially after 2-3 months of age).

### Table 1 - Synthesis with the series of cases discussed

<table>
<thead>
<tr>
<th>Case series</th>
<th>Symptomatology</th>
<th>Proposed treatment</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient 01:</td>
<td>Gait with increased base of support, hyperlordosis and difficulty abducting the hips.</td>
<td>Surgical correction in 2 steps: 1 - Open reduction, femoral shortening and Salter-type pelvic osteotomy on the right. 2 - After six months, the same procedure was performed on the left hip.</td>
<td>Good acetabular coverage on both hips. On the right, the head of the femur showed changes suggestive of avascular necrosis sequelae.</td>
</tr>
<tr>
<td>Patient 02:</td>
<td>Bilateral hip dislocation.</td>
<td>Conservative treatment using the Pavlik harness.</td>
<td>He presented a satisfactory response with good acetabular remodeling.</td>
</tr>
<tr>
<td>Patient 03:</td>
<td>Ortolani and Barlow were negative. In addition to hip USG within normal limits.</td>
<td>Outpatient follow-up.</td>
<td>Normal motor development.</td>
</tr>
</tbody>
</table>
The Barlow test is performed with the hip in flexion, adduction and neutral rotation. An attempt should be made to dislocate the hip by applying vertical force, from anterior to posterior. The patient’s thigh is at a right angle, the upper part of the femur is held between the index and middle fingers over the greater trochanter, and the thumb is held in the medial and proximal region of the thigh. The maneuver force will be exerted on the child’s knee in a vertical direction to the hip in an effort to displace the femoral head from within the acetabulum, in which the doctor looks for a “pistoning” sign in the hip, which may be accompanied by a “bounce”; the test is positive when the hip actually dislocates. In the 2nd phase of the test, the hip will be reduced with abduction and light medializing force performed with the index and middle fingers on the greater trochanter.

According to Vilate and Skaggs, the most common iatrogenic complication present among the different DDH treatments is osteonecrosis of the femoral head, resulting from blood supply interruption to the proximal epiphysis of the femur caused by compression or stretching of vessels in this region after concentric reduction.

Routine physical examinations from birth until the age at which the infant begins to walk is the best way to identify DDH early. The Barlow and Ortolani maneuvers are recommended for babies. According to Guarniero (2010, p. 118), the maneuvers need to be performed with the newborn calm, facilitating reduction. The hip abduction limitation becomes evident within a few weeks (in normal cases it exceeds 60 degrees). As the child grows, the Ortolani maneuver is no longer positive and the abduction limitation becomes more pronounced.

The Ortolani maneuver is recommended and performed throughout the world for all newborns. From this perspective, there is a discussion about when an USG should be requested in order to reduce non-diagnosis and late diagnoses of the disease. However, the preferred screening method for diagnosing DDH still remains controversial and radiographic examination is only indicated after 4 months of age.

In the United States, the Pediatric Orthopedic Society of North America (POSNA) recommends that a physical examination be carried out during the first months of life primarily by a pediatrician, and when positive, referral for a more detailed analysis with a pediatric orthopedist to confirm or reject the diagnosis. USG is performed for positive cases and those at high risk for DDH, with an agreement of 87.5% between the echogram and the positive result of the physical examination.

The newborn healthcare guide for orthopedic problems from the Ministry of Health (MoH) in Brazil recommends the Ortolani
maneuver in the first two days of life and in subsequent consultations\textsuperscript{4,15}. USG is recommended after a positive physical examination, heredity, oligohydramnios, breech presentation, congenital torticollis or foot malformations\textsuperscript{16}.

This change does not cause deformities in most cases, is painless and does not limit movement, meaning that the newborn is apparently normal. As there are no other significant clinical signs, dysplasia will only be diagnosed if objectively investigated\textsuperscript{16,17}.

According to Vaquero-Picado et al., the acetabular correction potential decreases drastically after 3 to 4 years of age. Early interventions are essential to obtain the best results with less surgical aggression. All treatments are based on seeking to position the femoral head concentrically in the acetabulum so that it is stimulated to grow normally\textsuperscript{10}.

According to Yang et al., a severe condition of non-reducible hip dislocation can present a negative result in the Barlow and Ortolani maneuvers. These maneuvers also depend on the technical competence of the professional, in addition to being more sensitive in newborns due to the greater laxity of the soft tissues around the hip joint\textsuperscript{11}. However, according to Rosenberg et al., the sensitivity of these two maneuvers separately is up to 54%, and therefore complementary imaging exams are appropriate for monitoring and/or diagnosis\textsuperscript{12}.

A useful way to evaluate hips with suspected dysplasia is ultrasound examination, especially in children under six months of age, when the proximal epiphysis of the femur and part of the acetabulum are cartilaginous. Hip analysis uses two methods described as morphological and static. The first is the dynamic method, created by Harcke, which studies joint mobility based on Barlow and Ortolani maneuvers. On the other hand, the second analyzes the proximal part of the femur and the pelvis contour presented by Graf\textsuperscript{19}.

Appropriate early neonatal diagnosis and treatment remain the best ways to achieve normal hip anatomy in children with non-tetralogical DDH\textsuperscript{19}. According to Ömeroğlu et al., early use of the Pavlik harness in children up to 3 months of age presents better results compared to children above that age. Ultrasound of the hip using the Graf method should be used for primary diagnosis and evaluation in the follow-up of DDH, showing that Graf type II hips have a higher success rate. Furthermore, it is considered therapeutic success when the Graf type I hip is achieved, because it has good bone and cartilaginous coverage of the femoral head\textsuperscript{20}.

In the prospective cohort study conducted by Gonzalez et al., 2 of the 34 newborns with suspected DDH had diagnostic confirmation on ultrasound. Both received conservative treatment using the Pavlik harness, observed for eight weeks, and had satisfactory results in both infants at the end of treatment. This result was also found in the second child, BLQ, with bilateral hip dislocation and submitted to the same therapy\textsuperscript{21}.

Treatment with the Pavlik harness fails in 2% of unstable hips and in up to 26% of patients with dislocated hips. When this happens, it is necessary to move on to a more invasive procedure, which may consist of adductor tenotomy and closed reduction of the hips, or open reductions via the Ludloff approach (medial) in younger children and anterior or anterolateral in older children\textsuperscript{12}.

Once the diagnosis of hip instability or dislocation is confirmed, treatment should be indicated as early as possible. This treatment in the first months of a newborn’s life seeks to reduce the femoral head in the acetabular cavity. Thus, by maintaining a reduced position of the hip in flexion, as well as in slight abduction, normal joint development will occur\textsuperscript{2}.

Treatment with the Pavlik harness can be performed up to the sixth month of life. Despite being simple, its application requires essential care. The anterior straps maintain hip flexion, which should remain at 90-100 degrees, as greater flexion can lead to femoral nerve paralysis. The posterior straps maintain abduction, which must be in a human position (the limbs naturally fall into abduction) and it must remain at a minimum of 60°, reducing the risk of avascular necrosis of the femoral head. In frankly dislocated hips, it is expected that correct use the hips will be stable within three to four weeks, and the harness must be maintained for at least eight weeks. Clinical and ultrasound control is performed weekly in the first four weeks and in the eighth week.

Although very efficient, the method can fail in 2% of unstable hips and in up to 26% of patients with dislocated hips. Other techniques are necessary when this happens, such as adductor tenotomy and closed reduction of the hips, or open reductions via the Ludloff approach (medial) in younger children and anterior or anterolateral in older children\textsuperscript{12}.

The Frejka appliance has been associated with poor outcomes and high rates of avascular necrosis because it has a tendency to force abduction of the hips. On the other hand, research shows that treatment with the Pavlik harness is superior in terms of successful reduction and has lower avascular necrosis rates\textsuperscript{22}. It is important to highlight that double diapers do not bring any benefit in treating DDH, as this is one of the points most questioned by parents during consultations\textsuperscript{8}.

Radiography can evaluate ossification and adequate development of the femoral head and acetabulum, as well as possible avascular necrosis. Several parameters have been used to evaluate hip development, among which the following stand out: the Menard-Shenton lines, which evaluate the relationship between the acetabulum and the femoral neck; the acetabular index, the Ombredane Quadrants (formed by the Perkins and Hilgenreiner lines, the secondary ossification nucleus of the femoral epiphysis must be located proximally in the inferomedial quadrant), useful for predicting the amount of concentrically reduced femoral head\textsuperscript{10} (Figure 9).

Yang et al. argues that the objective of treating DDH would be to reduce the incidence of total hip arthroplasty in adulthood due to non-treatment of DDH, prevent avascular necrosis of the femoral head and treat hip dysplasias and dislocations during childhood\textsuperscript{11}.

Barbosa and Albernaz consider hip ultrasound to be ideal for all female newborns, and the examination to be carried out in boys who present one of the following positive conditions, namely: positive family history of DDH, positive result for the Ortolani maneuver or pelvic presentation\textsuperscript{12}.

Goiano recommends diagnostic investigation in all maternity hospitals for newborns of both sexes, regardless of the presence or absence of risk factors\textsuperscript{1}.
CONCLUSION

An early and appropriate clinical evaluation by the physician should be performed on all newborns, regardless of sex. It is concluded that USG of the hips should be indicated for all newborns with the following conditions: positive family history of DDH (heredity), positive result for the Ortolani maneuver, breech presentation, oligohydramnios, congenital torticollis or foot malformations. Furthermore, early treatment in childhood is important, as it can prevent complications in adulthood and reduce the number of cases of early hip osteoarthritis.

Acknowledgements: We would like to thank LIGORTRN (Liga de Ortopedia e Traumatologia do Rio Grande do Norte) for their contribution to this study.

Financing: This work did not receive financing.

Conflict of interests: The authors declare there are no conflicts of interest.

Authors’ contributions: Vítor Ferraz Silva Tacconi - contributed to the study conception and planning, data analysis and interpretation, writing and critical review; and approved the final version. Tabata de Alcantara - Responsible for obtaining the data, contributed to the study conception and planning, data analysis and interpretation, writing and critical review; and approved the final version. João Paulo Araújo Dantas - contributed to the study conception and planning, data analysis and interpretation, writing and critical review; and approved the final version. Dayanna Bianca Bezerra de Mello - contributed to the study conception and planning, data analysis and interpretation, writing and critical review; and approved the final version. Àlex Espinalt Daví Lima de Freitas - contributed to the study conception and planning, data analysis and interpretation, writing and critical review; and approved the final version.

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