



# Profile of Patients Diagnosed with Developmental Dysplasia of the Hip\*

## *Perfil dos pacientes diagnosticados com displasia do desenvolvimento do quadril*

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### Abstract

**Objective** To describe the profile of patients with developmental dysplasia of the hip (DDH) diagnosed by physical and ultrasound examination, with the implementation of a protocol for the treatment and follow-up of DDH.

**Methods** A cross-sectional study with DDH patients born between January 2014 and December 2016, in the city of Pelotas, Southern Brazil. Ethnicity, gender, birth weight, fetal presentation, affected side of the hip, gestational age, maternal age and family history were considered. The data on the medical records were compared with the characteristics of the general population described on the Brazilian National Information System on Live Births (Sistema de Informação sobre Nascidos Vivos [SINASC]).

**Results** A total of 33 DDH patients were identified, mostly female, with a four-fold higher probability of having the condition ( $p < 0.001$ ); the left was the most affected side. No statistically significant association was found regarding the following factors: birth weight, gestational age, ethnicity, and maternal age. The newborns in breech presentation had a 15-fold higher probability of presenting DDH ( $p < 0.001$ ). A total of 21 newborns required immediate treatment of the hips, since the ultrasound showed a Graf classification of IIb or higher, or the radiography showed dislocation in DDH patients older than 6 months of age.

**Conclusion** Screening for DDH is essential in all newborns; physical examinations revealing alterations must be complemented with ultrasound imaging to avoid the delayed diagnosis of the condition.

### Keywords

- ▶ congenital hip dislocation
- ▶ developmental dysplasia of the hip
- ▶ neonatal screening

### Resumo

**Objetivo** Descrever o perfil dos pacientes com displasia do desenvolvimento do quadril (DDQ), diagnosticados por meio de exame físico e ultrassonográfico, com a implantação do protocolo de atenção e rastreamento de DDQ.

**Métodos** Estudo transversal que incluiu os portadores de DDQ nascidos de janeiro de 2014 a dezembro de 2016, na cidade de Pelotas, Sul do Brasil, que considerou os

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**Palavras-chave**

- ▶ luxação congênita de quadril
- ▶ displasia do desenvolvimento do quadril
- ▶ triagem neonatal

fatores etnia, sexo, peso ao nascer, posição fetal, lado de ocorrência, idade gestacional, idade materna e histórico familiar. Os dados de prontuário foram comparados com as características da população geral por meio do Sistema de Informação sobre Nascidos Vivos (Sinasc).

**Resultados** Foram identificados 33 portadores de DDQ, a maioria do sexo feminino, que mostrou uma probabilidade quatro vezes maior de apresentar a patologia, ( $p < 0,001$ ) e o lado mais acometido foi o esquerdo. Os recém-nascidos com apresentação pélvica tiveram uma probabilidade 15 vezes maior de ter DDQ ( $p < 0,001$ ). Não foi encontrada associação estatisticamente significativa com os seguintes fatores avaliados: peso ao nascer, idade gestacional, etnia e idade materna. Um total de 21 recém-nascidos necessitaram de tratamento imediato do quadril; a ecografia demonstrou classificação IIb ou maior, pelo método de Graf, ou a radiografia mostrou luxação nos portadores de DDQ com mais de seis meses de idade.

**Conclusão** O rastreio de DDQ é essencial em todos os recém-nascidos, e o exame físico, quando alterado, deve ser complementado com o ultrassonográfico para evitar o diagnóstico tardio da doença.

**Introduction**

Developmental dysplasia of the hip (DDH) is the term used to describe all changes in the hips of newborns, ranging from instability to dislocation. It replaced the previous term “congenital hip dislocation”, which, as implied, encompasses only dislocated hips.<sup>1</sup>

Currently, the screening for DDH in newborns employs the maneuver described by Ortolani<sup>24</sup> for the assessment of hip dislocation and instability; in some cases, the screening is performed through an ultrasound (US) examination using the method described by Graf, who classifies type I hips as mature and stable, type IIa hips as immature, type IIb hips as immature and unstable, type IIc hips as unstable, and types III and IV hips as dislocated.<sup>2,3</sup> When the physical or US examination is positive, the Pavlik brace, a flexible orthosis to keep the hips in from 90° to 110° of flexion and safety zone abduction, between 30° and 60°, is used.<sup>4</sup> The success rate of this treatment ranges from 86 to 99%.<sup>1,5,6</sup> These figures are easily explained by the fact that early reduction of the preserved neonatal hip anatomical structures results in normal joint growth.

Cases with late diagnosis and treatment are still the main cause of early hip arthritis, resulting in pain, functional disability and total hip arthroplasty in young adults.<sup>1</sup> Half of these patients with DDH with late diagnosis and treatment will present some degree of hip joint degeneration between the ages of 16 and 31 years old.<sup>7</sup>

The physical characteristics of the newborn and his/her mother may lead to a higher probability of DDH, increasing its prevalence in about 60%. The factors related to this increase are: female gender, white ethnicity, birth weight above 4,000 g, positive family history, gestational age over 40 weeks, primiparous mother, maternal age over 35 years old, feet morphological changes, and breech fetal presentation. The increase is even greater when these factors are associated.<sup>8-10</sup>

The purpose of the present study was to trace the profile of patients with DDH in the city of Pelotas, in the state of Rio Grande do Sul, Southern Brazil, and to implement a protocol for its screening.

**Material and Methods**

This is a cross-sectional, analytical study conducted at the only referral service for pediatric orthopedic treatment in the city of Pelotas, RS, Brazil. The medical records of DDH patients treated from January 2014 to December 2016 at the Children's Orthopedics Outpatient Clinic of our institution were evaluated; in addition, the authors prepared a questionnaire to assess the following variables described as associated with an increased DDH prevalence: ethnicity, gender, maternal age, parity, gestation time, birth weight, family history, fetal presentation and associated orthopedic malformations.

The characteristics of these newborns were compared to those of live births in Pelotas during the study period. These data were obtained from the Brazilian National Information System on Live Births (Sistema de Informação sobre Nascidos Vivos [SINASC]), since this information is routinely sent by maternity hospitals to the municipal Health Department. Patients with neural tube malformations, arthrogyrosis, and neuromuscular syndromes and diseases were excluded. The study was previously approved by the institutional Ethics in Research Committee under CAAE number 62063116.3.0000.5339.

A descriptive analysis of births in the city of Pelotas from January 2014 to December 2016 was performed using SINASC data, as well as a description of DDH cases treated at the referral service. The variables described in the SINASC and on the medical records were submitted to a bivariate analysis using the Chi-squared test or the Fisher test when indicated. The analyses were performed using the Epi Info 7.2. The level of statistical significance was set as 95%.

## Results

The characteristics of the births mentioned in the SINASC are shown in ►Table 1. From 2014 to 2016, 14,106 children were born in the city of Pelotas, an average of 4,702 births per year.

We found a total of 35 patients with DDH who were examined at the referral service. Two were excluded from the sample because they had not been born in Pelotas. Out of the remaining 33, 21 required immediate treatment of the hips, since their ultrasound Graft classification was IIb or higher, or the radiograph showed dislocation in infants older than six months. The other 12 patients were classified as IIa on the ultrasound examination; these infants were followed up at appointments held every four weeks, which included new physical and imaging examinations. The characteristics of the newborns with DDH are shown in ►Table 2.

The bivariate analysis between maternal and newborn characteristics and DDH showed a statistically significant

**Table 1** Profile of newborns from the city of Pelotas, RS, Brazil – SINASC<sup>a</sup> (2014 to 2016)

Characteristics	n (%)
<i>Gender</i>	
Male	7,185 (50.93)
Female	6,920 (49.06)
<i>Ethnicity<sup>b</sup></i>	
White	6,207 (79.63)
Mixed	489 (6.27)
Black	1,090 (13.98)
Other	04 (0.05)
<i>Weight</i>	
Lower than 3,000 g	4,802 (34.18)
3,000-3,999 g	8,564 (60.95)
Higher than 4,000 g	683 (4.86)
<i>Duration of pregnancy</i>	
Up to 37 weeks	2,064 (15.60)
37-41 weeks	1,0887 (82.29)
Over 41 weeks	279 (2.10)
<i>Maternal age</i>	
Up to 19 years old	2,042 (14.47)
19-34 years old	9,966 (70.65)
Over 35 years old	2,097 (14.86)
<i>Congenital abnormalities</i>	
Yes	98 (0.72)
No	12,492 (92.80)
Not informed	871 (6.47)
TOTAL	14,106 (100)

Notes: <sup>a</sup>Brazilian National Information System on Live Births (Sistema de Informação sobre Nascidos Vivos). <sup>b</sup>Data on ethnicity were not collected in 2016; for the remaining variables, the maximum percentage of unknown data was of 6.2%.

association regarding the child's gender. Girls were almost 4 times more likely to have DDH (prevalence ratio [PR] = 3.86; 95%confidence interval [CI] = 1.68–8.88;  $p < 0.001$ ). The variables birth weight ( $p = 0.90$ ), gestational age ( $p = 0.16$ ), ethnicity ( $p = 0.53$ ), and maternal age ( $p = 0.59$ ) had no statistically significant association with the outcome. Ethnicity was only evaluated for those born in 2014 and 2015, as this data was not included in the SINASC for the year 2016. Newborns in breech presentation were 15 times more likely to have DDH (PR = 15.30; 95%CI = 7.57–30.92;  $p < 0.001$ ).

Four children had had associated orthopedic anomalies, including congenital clubfoot and postural clubfoot; two had been born with knee retrocurvation.

## Discussion

One of the limitations of the present study was the small number of identified cases, which may make it difficult to test some associations. In addition, some children may not have been taken to the referral service because they were not properly examined at birth. Despite this potential selection bias, the occurrence of 2.3 cases per 1,000 births is similar to what is described in the literature. There is a huge variation in DDH incidence depending on ethnicity, habits, and the geographic region where the population lives. The incidence of neonatal hip joint dislocation and instability is of 2/1,000 and 10/1,000 births respectively.<sup>1</sup> Regarding ethnicity, the condition is uncommon among people of black ethnicity, and is highly prevalent among Native American (76/1,000) and Inuit (25-40/1,000) populations.<sup>11</sup>

Wynne-Davies<sup>12</sup> inferred that DDH results from the combination of instability due to increased hip joint laxity and environmental factors that lead to its onset. Since boys are less affected, it is believed that there is a hormonal cause or a less pronounced joint laxity in males.<sup>12,13</sup> The present study detected a higher prevalence in girls, a finding similar to those previously reported.<sup>2,9,11,13,14</sup>

It is known that between 60% and 93% of DDH patients present condition-associated factors.<sup>15-17</sup> The breech presentation would increase risk by 5-16 times.<sup>8,9,17</sup> In the present study, breech presentation was reported in 36% of infants with DDH, and it significantly increased the likelihood of the condition (15-fold higher risk). The literature indicates that family history increases the incidence of DDH from 12 to 68 cases per 1,000 births;<sup>3,6,18-20</sup> 24% of our sample had a previous history of the condition. Black ethnicity is mentioned as a protective factor, since DDH incidence in this population is three times lower compared to that of Caucasians;<sup>10</sup> however, this association was not confirmed in the sample evaluated in the present study. The association with birth weight was not statistically significant, although the literature indicates that birth weight is a risk factor when higher than 4,000 grams, and a protective factor if lower than 2,500 grams.<sup>8,9</sup> The SINASC had no information on parity, but, reportedly, the chance of first-term newborns having DDH is two to four times greater when compared to infants of mothers who already have two or more children.<sup>8,17,20</sup> This is due to the smaller size of the uterus at the first

**Table 2** Profile of newborns with developmental dysplasia of the hip (n = 33)

Characteristics %	
<i>Gender</i>	
Male	21.21
Female	78.78
<i>Ethnicity</i>	
White	69.69
Mixed	12.12
Black	9.09
Other	9.09
<i>Maternal age</i>	
Up to 19 years old	9.09
19-34 years old	69.69
Over 35 years old	15.15
Unknown	6.06
<i>First child</i>	
Yes	63.63
No	27.27
Not informed	9.09
<i>Duration of pregnancy</i>	
Up to 37 weeks	18.18
37-41 weeks	66.66
Over 41 weeks	6.06
Unknown	9.09
<i>Fetal presentation</i>	
Head	57.57
Breech	36.36
Unknown	6.06
<i>Family history</i>	
Positive	24.24
Negative	57.57
Unknown	18.18
<i>Affected side</i>	
Right	12.12
Left	57.57
Bilateral	21.21
Unknown	9.09
<i>Birth weight</i>	
Lower than 3,000 g	30.30
3,000-3,999 g	54.54
Higher than 4,000 g	6.06
Unknown	9.09
<i>Associated orthopedic abnormalities</i>	
Yes	72.72
No	12.12
Not informed	15.15
TOTAL	100.00

pregnancy.<sup>6,14,15,18,21</sup> In the present study, most cases were detected in first-born infants (64%).

Other factors described include the presence of oligohydramnios (four times more likely to occur in cases of DDH); in the present study, one case presented oligohydramnios<sup>6,8</sup> and postdatism.<sup>8,22</sup> According to the literature, maternal age is directly associated with DDH occurrence, which is twice as frequent when the mother is older than 35 years of age.<sup>8</sup> In the sample evaluated in the present study, age was not a factor that influenced the presence of DDH.

Associated malformations were identified in 12% of the sample, which is consistent with the findings by other authors.<sup>8,9,21,23</sup> The left side was the most affected (58%) in the present study, a finding that is once more in line with those of the literature.<sup>9,15,18</sup>

Screening for DDH, a condition that can cause pain, and functional and labor limitations when left untreated, has been performed for a long time. In 1937, Italian pediatrician Marino Ortolani<sup>24</sup> described the maneuver, which consists of hip flexion and abduction. In case the hip was dislocated, there was a bounce, since the described movement reduces the dislocation.<sup>21,24</sup> In 1957, Von Rosen<sup>25</sup> introduced the routine screening of all newborns with the Ortolani maneuver in Sweden, drastically reducing DHH cases with delayed diagnosis.<sup>21,25</sup> Today, the Ortolani maneuver is unanimously performed worldwide, since it is indicated to all newborns. The discussion is regarding the time when a complementary imaging test (ultrasound) should be performed to minimize the delayed detection and undiagnosed cases of DDH.<sup>26</sup>

The treatment has a 90% success rate in cases of early diagnosis, before six months of life, combined with the use of Pavlik braces.<sup>6</sup> This is because the hip joint of the newborn, although dislocated, is not morphologically altered, and the orthosis can reduce and stabilize it, mostly resulting in normal development.

The complication rate with braces is low, less than 1%. The major treatment complication is avascular necrosis of the femoral epiphysis. Femoral nerve palsy may also occur, with complete remission after brace removal.<sup>14</sup> Undiagnosed cases or cases with delayed diagnosis after gait initiation (at 12 months old) lead to early joint degeneration, with 86% of surgical indication in cases that are undiagnosed until 10 months of age.<sup>7,22</sup> In addition, it is known that 25 to 40% of early cases of hip osteoarthritis are secondary to cases of DDH that were neglected or submitted to delayed treatment.<sup>27</sup> In cases of delayed treatment requiring surgery (pelvic or femoral osteotomy), 44% will have some degree of arthrosis between the ages of 16 and 31 years.<sup>7</sup> At 33 years after treatment of DDH with late diagnosis, 50% of the cases will develop moderate or severe arthritis, and 14% will probably have been submitted to a total hip arthroplasty (THA), and, at 45 years after treatment, 54% will have undergone THA, and a third of the remaining cases will develop coxarthrosis.<sup>15</sup>

The preferred screening method for DDH diagnosis remains controversial. Currently, the screening methods include physical examination and US. The use of radiographic examination is only indicated in children older than four months.<sup>18</sup>

Mahan et al<sup>6</sup> described three DDH screening models: the first is the universal physical examination (Ortolani and Barlow maneuvers); the second is the US in cases of positive physical examination, breech presentation and family history; and the third model is the universal US, to which all newborns would be submitted. Their study showed that the chance of having early hip degeneration with consequent arthritis was higher in cases not submitted to US compared with the other two US models. When comparing universal US with positive physical examination or risk factors, they noticed an increase in false-positive diagnoses regarding the first model. However, the incidence of late cases or early joint degeneration did not change in both groups.<sup>6</sup> Schams et al<sup>3</sup> recommend the universal model, since the combination of two risk factors showed no evidence of a higher risk of presenting DDH. Bache et al<sup>14</sup> recommend US in all female newborns and in male newborns with a risk factor for DDH, whereas Woodacre et al<sup>17</sup> observed that family history and breech delivery or presentation were the only risk factors associated with DDH in males.

As recommended by the Pediatric Orthopaedic Society of North America (POSNA), the physical examination is critical; it must be performed by a pediatrician and, if positive, the case must be referred to an orthopedist to clarify or confirm the diagnosis of DDH. Thus, the physical examination is superior to imaging (US).<sup>26,28</sup> An US examination must be performed on hips at high risk for DDH or in cases of positive physical examination. The rate of agreement between the physical examination and the US is of 87.5%.<sup>27</sup> It is known that 60 to 80% of newborn hip abnormalities detected by physical examination resolve within 2 to 8 weeks, as do 90% of newborn hip abnormalities found through US.<sup>14,26</sup>

In Brazil, the Ministry of Health (MH) recommends the Ortolani maneuver in the first two days of life and subsequent childcare consultations, while US is recommended when the Ortolani maneuver is positive or in the presence of family history, breech presentation, congenital torticollis or feet malformations.<sup>29</sup>

## Conclusion

Although the screening method considered ideal by the present study is hip US in all female newborns and in male newborns with one of three characteristics (positive Ortolani maneuver, breech presentation and family history), in the city of Pelotas, as well as in most regions of Brazil, this model is not feasible due to the cost, logistics and difficulty to obtain specialized professionals and equipment to meet the demand created by it. The recommendation is to effectively follow the MS protocol to avoid late cases of DDH in our country. All maternity centers must follow the MH guidelines: Ortolani maneuver in the first two days of life and in subsequent consultations, and US when the maneuver is positive or in the presence of family history, breech presentation, congenital torticollis or feet malformations.

## Conflicts of Interest

The authors have none to declare.

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