Original Article

Therapies for neonates with congenital malformations admitted to a neonatal unit

Terapêuticas utilizadas em recém-nascidos com malformações congênitas internados em unidade neonatal

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ABSTRACT

The aim of this study was to characterize the treatments applied to newborns with congenital malformation hospitalized in a neonatal unit and to identify whether there is an association among the treatments used and the type of malformation. A descriptive, prospective and quantitative study was developed in a public institution in Fortaleza, Ceará, Brazil. Data were collected using the medical records of 30 neonates with congenital malformations. The incidence of malformations was higher among females, regardless of the mother's age, gestational age or weight at birth; malformations of the central nervous and musculoskeletal systems prevailed. The treatments used varied according to the clinical evolution of the neonate. The data collected did not present statistical significance when associated with the variable of congenital malformation and the treatments used ($p>0.05$). The treatments are not directly related to the type of malformation, but to the clinical condition of the neonate.

Descriptors: Infant, Newborn; Congenital Abnormalities; Neonatal Nursing; Therapeutics.

RESUMO

Objetivou-se caracterizar as terapêuticas aplicadas aos recém-nascidos com malformações congênitas internados em Unidade Neonatal e identificar se existe associação entre as terapêuticas utilizadas e o tipo de malformação. Estudo descritivo, prospectivo, quantitativo realizado em instituição pública de Fortaleza - CE, Brasil. A coleta de dados foi realizada por meio dos registros dos prontuários de 30 neonatos com malformações congênitas. Verificou-se que a ocorrência de malformações foi maior no sexo feminino, independente da idade materna, idade gestacional ou peso ao nascer; prevaleceram as malformações do sistema nervoso central e osteomuscular. As terapêuticas utilizadas variaram conforme a evolução clínica do neonato. Os dados não demonstraram significância estatística quando associada à variável malformação congênita e às terapêuticas utilizadas ($p>0,05$). As terapêuticas não estão diretamente relacionadas ao tipo de malformação, mas ao quadro clínico do neonato.

Descritores: Recém-nascido; Anormalidades Congênitas; Enfermagem Neonatal; Terapêutica.

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INTRODUCTION

Congenital malformations may be defined as any functional or structural anomalies in fetal development, due to factors originating before birth, of genetic, environmental or unknown nature, even when the defects are not apparent in neonates (NNs) and appear later[1].

Several internal or external factors may contribute to the development of genetic defects; however, the causes are unknown in most cases. Of particular note, besides mortality, malformations also account for high rates of morbidity and risks for the development of clinical complications, with numerous admissions and serious complications[2]. Every year, around 7.9 million neonates worldwide – 6% of all births – are born with serious congenital defects. Among these, almost 3.2 million are permanently disabled, requiring clinical follow-up[3]. Populations are exposed to the risks of congenital malformations, but frequency and type vary by race, ethnic group and socioeconomic conditions[4].

For better organization of existing congenital malformation categories, the tenth revision of the International Classification of Diseases was conducted by the World Health Organization (WHO), resulting in the following categories: congenital malformations of the eye, ear, face and neck; congenital malformations of the circulatory system; congenital malformations of the respiratory system; cleft lip and/or cleft palate; other congenital malformations of the digestive system; congenital malformations of the genital organs; congenital malformations of the urinary system; congenital malformations and deformations of the musculoskeletal system; other congenital malformations; and chromosomal abnormalities[5].

The birth of preterm neonates (PTNNs) or neonates with any congenital malformations requires admission to units with specialized technological, human and therapeutic resources that ensure more complex care. Such support is found in neonatal intensive care units (NICUs) and is provided continuously, which has contributed to longer survival of preterm neonates, especially those presenting extremely low weight[6].

Neonatal intensive care units are formal units created, organized and based on professional knowledge that requires specificity, ability and principles focused on PTNNs or NNs with any diseases, and malformed NNs are inserted in this context[7]. Longer survival of neonates with malformations has significantly improved in recent years, supported by progress in surgical techniques and new therapeutic resources, which has contributed to eliminating surgically correctable anomalies[8].

Hospitalization and, consequently, NICUs are associated with the fact that neonates are submitted to an excessive number of procedures, such as venous puncture, orogastric and vesical intubation, capillary glycemia, dressings, pulmonary aspiration, endotracheal intubation, and drain removal, among others, which may cause discomfort, stress and pain [6,9].

Based on the considerations above, this study is relevant as it contributes to individual neonatal nursing support practice, with detailed information about therapy used during hospitalization of neonates with congenital malformations, and increases knowledge of healthcare professionals about this specific group of patients, supporting additional studies and enhancing nursing care to neonates with malformations.

Knowing that malformed neonates, once hospitalized in these units, will require therapies to keep health integrity, the following questions were asked: What therapies are implemented in neonatal units for neonates born with congenital malformations? Is there any association between neonatal therapies and the types of malformations presented, according to the ICD-10 classification?

Thus, the purpose of this study was to characterize the therapies applied to neonates with congenital malformations admitted to a neonatal intensive care unit and identify any associations between therapy and malformation type.

METHODS

An exploratory, descriptive, prospective and quantitative study. It is an excerpt of a larger study titled "Neonates admitted to a neonatal unit: emphasis on pain evaluation and congenital malformation prevalence" (*Recém-nascidos internados na unidade neonatal: ênfase na avaliação da dor e na prevalência da malformação congênita),* which was supported by the Brazilian Council for Scientific and Technological Development (CNPq), according to process 483352/2011-0. It was conducted in neonate hospitalization units 1 and 2 in a public institution in the city of Fortaleza, Ceará, Brazil, a reference center in the state. The sectors had 54 beds, 21 for high-risk patients and the remaining beds for medium-risk patients.

The studied population included all neonates who presented any type of congenital malformations at birth, apparent or not, during the data collection period from November 2011 to January 2012, and who were sent to the high- or medium-risk neonatal unit, resulting in a sample of 30 neonates.

The study included neonates with any congenital anomalies admitted to the neonatal unit who were born in the studied institution or transferred from other institutions during the data collection period, whose weight was above 500 g, regardless of gestational age, gender, length, number of gestations, type of childbirth, Apgar score, color, diagnosis, oxygen therapy, medication therapy and nutrition therapy.

The exclusion criterion was malformed neonates with death confirmed immediately after birth, as evaluated by the neonatologist in the childbirth room, without requiring transfer to the NICU.

Data were collected from clinical records of neonates with congenital malformations, using a form with variables like maternal age, gestational age, gender, weight, and type of congenital malformation. The form had other variables that characterized the following therapies: ventilation type, nutrition therapy, medication therapy, dressing, venous access, intervals to control water balance, and surgery.

These data were organized, categorized and evaluated according to the variable type using Microsoft Office Excel and analyzed using the Statistical Package for the Social Sciences (SPSS®) version 20. Bivariate analysis was used to evaluate the association between the malformation categories and the variables related to oxygen therapy, nutrition, water balance, dressing (site and type), surgery, venous access and medication therapy.

Descriptive statistics with simple and absolute frequency and dispersion measurements such as mean and standard deviation were applied as appropriate. For inferential statistics, the chi-square test was used to check for any association of the categorical variables and the Spearman's correlation coefficient was used for quantitative variables; a significance level of 5% ($p<0.05$) was considered for both.

For classification of congenital malformations, the International Statistical Classification of Diseases and Related Health Problems, tenth revision (ICD-10) was used, which provides disease codes and a variety of related signs and symptoms.

The project was approved by the institution's Ethics Committee under protocol number 546/2011, respecting the National Health Council guidelines on ethical aspects of research with human beings, as defined in Resolution 196/96. The parents or guardians of malformed neonates signed an informed consent form authorizing the participation of the neonates in the study.

RESULTS

To better describe the results, tables were developed to list relevant variables in the study and their values, in accordance with the tests conducted with the studied sample.

Table 1 Profile of neonates with congenital malformations. Fortaleza, CE, Brazil, 2012.

Variables	N	%	Mean	SD
Gender				
Female	19	63.3		
Male	11	36.7		
Weight at birth (grams)			2,726	985.4
<999	2	6.6		
1000 – 1499	1	3.4		
1500 – 2499	11	36.7		
2500 – 3999	13	43.3		
>4000	3	10.0		
Gestational age (weeks)			36.7	2.7
<30	1	3.4		
30-34	7	23.3		
35-36.9	7	23.3		
37- 42	15	50.0		
Type of childbirth				
Cesarean	18.0	60.0		
Vaginal	12.0	40.0		
Maternal age (years)				
≤19	60	20.0		
20 – 30	180	60.0		
31 – 40	60	20.0		

Source: Clinical records of neonates

Thirty neonates were analyzed, with a prevalence of females (19, 63.3%); gestational age rangied from 31 to 42 weeks (mean 36.7±2.7). The mean weight at birth was 2,726 g.

Maternal age varied from 19 to 40 years, but most were 20 to 30 years old (18, 60%, mean 24.6). Cesarean was present in 18 NNs, or 60% of the studied sample.

According to the ICD-10 classification, 48 congenital malformations were observed in 29 neonates, alone or in combination. Micrognathia was observed in one neonate; although classified as a malformation in the clinical records, it not included in Chapter 17 of the ICD-10, and for this reason, it is not in Table 2. Since it was a single case, it was not excluded from the study, resulting in a sample of 30 neonates. The most frequent cases were: 18 cases (37.5%) of total malformations of the musculoskeletal system, with predominance of congenital club foot (33.3%) and other malformations (22.2%), including reduction defects of the limbs, absence of fibula, and asymmetric skull; 11 (22.9%) malformations of the central nervous system (CNS), with myelomeningocele (45.4%) as the most frequent malformation, followed by hydrocephalus (36.4%); and cleft lip and/or cleft palate, observed in 5 cases. Less frequent malformations were in the respiratory system and chromosomal abnormalities, both 4.1% among total diagnosed cases.

Tables 3 and 4 show the therapies applied to neonates in the NICU, as well as their diversity and frequency of application.

Table 2: Categories of congenital malformations presented by neonates admitted to the neonate unit. Fortaleza, CE, Brazil, 2012.

Categories of congenital malformations	N	%
Musculoskeletal system (n=18)		
Congenital club foot	5.0	27.8
Other	4.0	22.2
Gastroschisis	3.0	16.7
Omphalocele	2.0	11.1
Diaphragmatic hernia	2.0	11.1
Macrocephaly	2.0	11.1
Central nervous system (n=11)		
Myelomeningocele	5.0	45,4
Hydrocephalus	4.0	36.4
Spina bifida with hydrocephalus	1.0	9.1
Encephalocele	1.0	9.1
Cleft lip and/or cleft palate (n=5)	5.0	100.0
Circulatory system (n=4)		
Malformation of tricuspid valve	1.0	25.0
Single ventricle	1.0	25.0
Ventriculomegaly	1.0	25.0
Cardiopathy	1.0	25.0
Eye, ear, face and neck (n=5)		
Ear dysplasia	2.0	40.0
Finger deformities	1.0	20.0
Malformation of outer ear	1.0	20.0
Macroglossia	1.0	20.0
Other congenital malformations of the digestive system (n=3)		
Atresia of esophagus	2.0	66.7
Atresia of duodenum	1.0	33.3
Chromosomal abnormalities (n=2)		
Down syndrome	2.0	100.0

Source: Clinical records of neonates.

Table 3: Therapies applied to neonates with congenital malformations. Fortaleza, CE, Brazil, 2012.

Variables (n=30)	N	%
Oxygen therapy		
Ambient air	14.0	46.7
Mechanical ventilation	6	20.0
Oxygen hood	6	20.0
CPAP*	4	13.3
Medication therapy		
Antibiotics	15	50.0
No medication	7	23.3
Antibiotics + fluid replacement	3	10.0
Fluid replacement	2	6.7
Opioids	1	3.3
Fluid replacement + oral medication	1	3.3
Antibiotics + opioids + fluid replacement	1	3.3
Nutrition		
Diet via orograstric tube	11	36.7
Parenteral nutrition	8	26.7
Oral diet	7	23.3
Zero diet	4	13.3
Surgery		
No	19	63.3
Awaiting	7	23.3
Yes	4	13.3
Water balance interval		
Every 3 hours	23	76.7
Every 2 hours	7	23.3

Source: Clinical records of neonates.

* Continuous positive airway pressure

Table 4: Therapies for dressings applied to neonates with congenital malformations. Fortaleza, CE, Brazil, 2012.

Variables (n=30)	N	%
Use of dressing		
No	19.0	63.3
Yes	11.0	36.7
Dressing site (n=11)		
Sacral area	6.0	54.5
Umbilical scar	3.0	27.3
Abdominal area	2.0	18.2
Type of dressing (n=11)		
Gauze + collagenase+ saline solution 0.9%	3.0	27.3
Sterile cover	2.0	18.2
No specification	2.0	18.2
Gauze + collagenase	1.0	9.1
Silver sulf. 1%	1.0	9.1
Gauze+ silver sulf. 1% + alcohol 70%	1.0	9.1
Gauze	1.0	9.1

Source: Clinical records of neonates.

Oxygen therapy was applied to 53.3% of the neonates. Regarding medication therapy, the predominance of antibiotics alone was 50% (15); 10% (3) received antibiotics combined with fluid replacement; 3.3% (1) received antibiotics with fluid replacement and opioids; and 23.3% (7) did not receive any medications. The others received therapy with opioids, oral medication or fluid replacement alone.

Regarding nutrition therapy, force feeding via orogastric tube predominated at 36.7% (11), followed by parenteral nutrition at 26.7% (8).

As for surgery, 63.3% (19) of the studied sample was not submitted to surgery and/or did not require surgery; however, 13.3% (4) were submitted to surgical procedure for congenital malformations during the data collection period.

Regarding the frequency of water balance, 76.7% (23) of neonates were handled every 3 hours to provide rigorous support for maintenance of hemodynamic balance.

Regarding the use of dressings, 11(36.7%) of the neonates received dressings,. The others did not receive specific dressings related to the diagnosis, but other devices were used, such as central venous catheters, with protection film adhered to the skin, which were not considered in this study.

Among the 11 dressings observed, most were located in the sacral region, 54.5% (6) of the cases, followed by the umbilical scar, 27.3% (3). The most frequent type of dressing was gauze combined with collagenase and saline solution 0.9%, accounting for 27.3% (3) of the dressings.

When investigating possible associations between malformations and the variables oxygen therapy, nutrition therapy, medication therapy, venous access type, water balance interval, surgery, and dressings (site and type), the following values were obtained: p=0.527; p=0.653; p=0.258; p=0.610; p= 0.400; p= 0.416; p= 0.399; p= 278; p=151, respectively. Thus, no statistically significant association was observed, with p >0.05.

DISCUSSION

In previous research, congenital malformations have occurred more often in males (in more than 50% of samples), which disagrees with the present study, where female neonates were predominant (63.3%)[2,10].

Regarding weight at birth, the 2,500 to 3,999 g range, considered the normal weight range, was observed in 43.3% of the sample, and neonates presenting weight lower than 2,500 g accounted for 46.7% of the sample; mean weight at birth was 2,726g±985.45. One study of birth weight weight found 10 neonates that weighed <2,500 g and 45 that weight ≥2,500 g, showing a higher frequency of normal weight among neonates[11].

However, divergences appear when stating that genetic or environmental factors may be the cause of congenital malformations and involved in low weight at birth, thus explaining the association between the presence of congenital malformations and low weight at birth [12].

Regarding gestational age, the majority of malformations occurred between weeks 37 and 41 (50% of the sample). A study in Turkey with a sample of 242 children that analyzed only malformations of the gastrointestinal system and congenital heart diseases, whose purpose was to investigate the relation between congenital heart defects and incidence and mortality of children with malformation of the gastrointestinal system, agreed with the present study in terms of mean gestational age of 37.2±2.6 and weight at birth of 2,756±612g[13]. A study conducted in Campina Grande, in the northeast region of Brazil, observed 190 congenital malformations from January 2003 to December 2005 and showed that most mothers presented gestational ages between 37 and 41 weeks and did not show statistical associations between gestational age and presence of congenital malformations (χ^2=0.57, p=0.30)[14].

It is important to analyze the statistical values of scientific evidence, not just for Brazil, but also at the global level, to allow relevant conclusions about the profile of malformed neonates.

A study conducted in Lebanon to evaluate the incidence, types and correlations of congenital malformations in Lebanese hospitals indicated a profile for neonates as follows: males were more prevalent, accounting for 58.3% of the sample, and 41.7% were females; mean weight at birth was 2,908.95±584.02; maternal age was 28.74±6.30; Apgar score at 5 minutes was 5.71±3.63, with weight and Apgar score presenting statistically significant relationships with congenital malformation, p=0.027 and p<0.001, respectively[15].

Regarding maternal age, the present study showed a prevalence of mothers aged between 20 and 30 years (60%). However, no statistical significance was observed in relation to malformations. Another study showed 26.4% of the mothers were adolescents, 60.2% adults and 13.4% late mothers. The chance of multiparous adolescents having children with congenital malformations was 6.14 times higher than for uniparous adolescents. However, the incidence in late mothers was 11.4 times higher, when compared to mothers between 20 and 34 years old[16].

In the present study, the most prevalent malformations in neonates were of the musculoskeletal system (17) and the CNS (11). The literature shows similar results in terms of prevalence of CNS and musculoskeletal malformations, at 31%[2]. Another study showed high rates of malformations in the nervous and musculoskeletal systems, 47.3% and 13.2%, respectively, followed by cleft lip and/or cleft palate at 3.5%[17].

Although these malformations present high percentages, those with lower impairment and complexity have a better prognosis, while the most complex malformations require outpatient care and interventions for functional recovery or reduced sequelae. The diagnosis of malformations of the musculoskeletal system may occur while in utero, with the support of ultrasound imaging. However, detailed evaluation with ultrasound is required so that minor anomalies whose clinical evaluation may indicate normal results do not go unnoticed [18].

Anomalies of the CNS, the second most common malformation category among the diagnoses in the present study, involve defects in the whole body, depending on the sites where they occur, since they play a role in organ development and organic functionalities.

In the present study, the classification that involved the CNS reached 22.4%. Similar data were obtained in a study conducted in Recife with 3,908 living neonates, in which the most prevalent anomalies were of the CNS (27.4%) and the musculoskeletal system (21.2%)[16].

It was not possible to analyze family history of congenital malformations, which did not allow correlation between prevalence of malformations and risk factors. In

addition, cases of fetal deaths or stillbirths due to malformations were not analyzed, because they were not the focus of this study.

Regarding therapies used with malformed neonates, they varied according to malformation type, neonate needs and risks the neonates were exposed to. The care provided to neonates with individual anomalies is not necessarily the same as for neonates born with congenital defects. For effective humanized service in neonatal units, integrated teams are required, in which the articulation of actions and knowledge among various professionals can provide support to neonates, improving service quality[19].

Oxygen therapy is used with all neonates requiring ventilation in their adaptation to extrauterine life[20]. In the present study, oxygen therapy was also administered to neonates with malformations directly related to the respiratory or digestive systems due to impaired spontaneous ventilation. In cases of cleft palate, the therapies were related to diet and ventilation, according to the neonate's needs.

For the survival of extremely low-weight and critically ill neonates, such as malformed neonates, nutrition support is essential to ensure growth and development[21]. In the present study, the care involved the prevention of complications these neonates could develop and in their clinical condition.

The most common procedure for cleft palate is surgical correction, conducted in one or more stages and outpatient follow-up. In cases of malformations, like gastroschisis and myelomeningocele, surgery is suggested within the first 24 hours of life, in an attempt to prevent infections or complications from the malformations, leading to reduced time in oxygen therapy and total parenteral nutrition and reduced hospitalization periods[22-23].

In terms of dressing, this procedure is required in certain malformations presenting significant and apparent lesions, like gastroschisis, omphalocele, and spina bifida, and particularly for lesions due to myelomeningocele. Regarding the results obtained, the most common dressing site was the sacral region, which was closely related to malformations of spina bifida.

In general, myelomeningocele, the most common malformation of the CNS according to the results, is located in the lumbrosacral region of the spinal column. The cerebrospinal fluid may flow through the defect, which is often covered by a fine membrane; otherwise, the tissue is exposed [22].

Performing nursing care for lesions due to myelomeningocele requires pressure control over body areas, observing lesion characteristics, applying a proper solution to the skin/lesion as appropriate, and providing adequate dressings[24]. Dressings for these lesions varies according to the neonatal unit routine in hospitals and the availability of proper materials for the lesion characteristics.

Therefore, through the diagnosis of malformations, sophisticated therapies, surgical procedures, modern appliances and devices and specialized professionals, a chance of life is ensured for neonates who, until some decades ago, were considered unviable by science[25].

CONCLUSION

Some malformations require specific care and therapies, according to the clinical progress of neonates and considering the anomalies presented, but they are not directly related to the type of malformation. However, the clinical situation of such neonates may be similar to that of other neonates without congenital defects.

In the present study, the most common therapies included oxygen therapy, antibiotic therapy, nutrition therapy via orogastric tube and dressing with collagenase, but they did not present statistically significant associations with congenital malformations.

Therefore, additional studies with larger samples are required to evaluate possible associations among the therapies used with neonates and congenital malformations. Based on scientific evidence, this study

attempts to provide nursing professionals and the academic community with knowledge of hospital practice with neonates with special needs, in order to improve support that is systematically and individually developed, ensuring better quality of life and promoting health among neonates in neonatal intensive care units, considering that most studies published to date have focused on the prevalence of malformations, not specifically considering nursing support for this specific population.

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