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NUTRITIONAL ASSESSMENT OF CHILDREN AND TEENAGERS WITH DOWN SYNDROME AND CONGENITAL CARDIOPATHY

EVALUACIÓN NUTRICIONAL DE NIÑOS Y ADOLESCENTES CON SÍNDROME DE DOWN Y CARDIOPATÍA CONGÉNITA

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ABSTRACT

The aim of this study was to analyze the nutritional status of patients with Down Syndrome (DS) and congenital heart defect (CHD). A transversal study which assessed the nutritional profile of DS and CHD patients between 0 and 18 years of age was done. Weight and stature, both at birth and current were considered. A total of 98 patients, 56% female were analyzed. Prevalence of low weight, overweight and low stature at the current anthropometric data (CAD) was 30%, 22% and 17%, respectively. There was a statistically significant difference between malnutrition at birth and the CAD ($p<0.01$). The current weight/age (W/A) ratio showed significant increase as compared to birth W/A ($p<0.05$). In addition, there was also stature recovery based on stature/age (S/A) ratio ($p<0,001$). In conclusion, at birth, these patients present stature and weight deficit but as age advances there is a stature improvement; however it is not properly accompanied by the weight, since there is a tendency towards overweight in puberty.

Key words: Down syndrome; congenital heart defect; nutrition assessment; obesity; malnutrition.

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INTRODUCTION

Down Syndrome (DS) results from trisomy of human chromosome 21, which determines, among other characteristics, mental and growth retardation (1-3). DS has an annual incidence of 1/800 living births worldwide and 8 thousand births in Brazil (3-5). Maternal age above 35 is one of the main risk factors for the development of such genetic defects (6). The main causes of death in DS patients are congenital cardiac complications. Nevertheless, thanks to medical advances both in therapeutics and diagnostics, life expectancy of such patients has increased dramatically over the last decades (4,7-9).

Approximately 40% of children with DS have congenital cardiopathies, the most frequent congenital malformation linked to DS (9). Half of those children present Atrioventricular Septal Defect (AVSD), which is much rarer in normal individuals. Other congenital cardiopathies such as Interatrial Communication (IAC), Interventricular Communication (IVC) and Patent Ar-

terial Duct (PAD) are also more frequent in DS patients (8-11).

Evaluation of nutritional status is usually performed through anthropometry. Special characteristics of DS patients, which are inherent to their condition, are considered in the curves suggested by Cronk and collaborators (7,12). These curves were elaborated based on the American population data and consider morbidities as hypothyroidism and congenital cardiac diseases (2,13,14).

It is known that weight and stature are significantly lower in children with cardiopathy, and low stature is one of the main characteristics of people with DS (4, 5, 10-12).

Nutritional assessment studies of DS patients are scarce in the literature (2). Therefore, it is extremely important to develop such studies, which will provide health care professionals with suitable tools and information that can adapt to the features of their target

population (2,16).

The objective of this study was to analyze the nutritional profile of children with DS and congenital cardiopathy from a public pediatric ambulatory specialized in cardiology, of the unified public Health Care System, in Porto Alegre – RS, Brazil.

SUBJECTS AND METHODS

This is a cross-sectional study in which ambulatory patients with DS and congenital cardiopathy, between 0 and 18 years (both male and female) were assessed. They were all admitted at the public pediatric ambulatory of Instituto de Cardiologia, Porto Alegre – RS, between January and July of 2010.

The project was approved by the Ethic Committee of the Instituto de Cardiologia do Rio Grande do Sul under authorization number 4389/09. After a detailed explanation on the study, it was handed and addressed to the patient's legal guardian an informed free consent term. The study included only patients whose legal guardian consented their participation by agreeing and signing the term.

The responsible relatives answered a questionnaire with questions on their educational level, pregnancy period, previous cardiac surgical procedure, type of cardiopathy, anthropometrical data of the child (or teenager) at birth as well as of today.

The nutritional evaluation of the patients was performed by the researchers in an objective way as follows. The measurement of the weight of children up to 2 years of age was done using a Balmak mechanical weighing scale with capacity of maximum 16 kg and 5 g accuracy. Children above 2 years of age were weighed on a Welmy electronic weighing scale with capacity of maximum 150 kg and accuracy of 50 g. Children were barefoot and with light clothing during weighing, according to international standards (17).

The stature of the children up to 2 of age was obtained with the use of an infantometer with accuracy of 0.1 cm, having the child lying stretched over a plain surface. For children older than 2 years of age, a stadiometer with accuracy of 0.1 cm was used, having the patient in vertical position, legs together and barefoot.

Children ages were calculated by taking the date on which they had their stature and weight measured and subtracting it by the date obtained on their vaccination cards, birth certificate and / or informed by their responsible relative.

In order to classify the nutritional status of the studied individuals, the weight/age (W/A) and stature/age (S/A) ratios were used and expressed in comparison to the median of the reference population (12). The cut-off

point for low W/A was percentile values below 10th (for children up to 3 years of age) and below 5th (for those older than 3 years of age). For overweight, percentile values above 95th were used. The cut-off points for low S/A were percentiles lower than 10th (for those up to 3 years of age) and lower than 5th (for those older than 3 years of age).

The data was stored and analyzed using the statistical package software SPSS version 18.0. The qualitative variables were obtained from absolute and relative frequencies. In order to compare the patient's nutritional status, both at birth and current, for weight and stature, Wilcoxon non-parametric t tests were used. To evaluate the relation between malnutrition and the qualitative variables, the Chi-Square test was utilized. The relation between the presence or absence of birth and current malnutrition, considering weight and stature, was done by using McNemar non-parametric test. The significance level adopted was of 5%.

RESULTS

Ninety-eight patients from the public pediatric ambulatory of Instituto de Cardiologia de Porto Alegre – RS were evaluated. The studied population was composed by 56.1% girls (n=55) and 43.9% boys (n=43), with ages between 0 and 18. The most frequent age group was the one with individuals between 10 and 18 years of age, (40.8%) (n=40), while the least frequent was of children between 6 and 10 years of age, representing 11.2% (n=11) of the studied population. The gestational time was predominantly of term births (≥ 38 weeks), (77.6%).

Most of the mothers, 66.3% (n=65), were older than 35 at the child's birth. In terms of maternal education, 61.2% (n= 60) had attended primary school, however only 28.3% (n=17) of these completed this initial educational level. The rest, 71.7% (n= 43), have not completed their studies. Mothers with higher education represented 7.2% (n=7), being 42.9% of these (n=3) incomplete and 57.1% (n=4) completed degrees.

Of the evaluated patients, 50% (n=49) had already undergone some cardiac surgical procedure, most of them (39%, n=38) before turning 2 years of age. There was no association between the type of cardiopathy and the nutritional status ($p>0,05$). Of the different congenital cardiopathies found in the studied population 50% (n=49) of the individuals presented AVSD, 28% (n=27) IVC, 24% (n=23) with problems in the IAC, 22% (n=21) with a flaw PAD and 13% (n=13) with Tetralogy of Fallot (TOF) (table 1).

As for the nutritional status at birth, it could be observed from the W/A curve that 31% (n=30) of the individuals presented low weight for their ages. Accord-

ding to the S/A curve, 42% (n=41) of the children were born with stature below normality.

Of the 98 patients, 52% (n=51) had difficulty to increase put on weight in the first months following birth, while only 26% (n=26) of them had adequate weight gain at this phase; the responsible relatives of the remaining patients 22% (n=21) were not able to report regarding weight gain of their children in the first

months following birth.

However, in the current nutritional status evaluation, a tendency towards gain weight by these individuals could be noticed. Overweight, based on the W/A, has been diagnosed in 22% (n=22) of the studied population. The number of evaluated patients with low weight was the same found at birth age, 31% (n=30). Regarding the S/A curve, 17% (n=17) of the cases presented low stature

TABLE 1

General characteristics of the 98 children and teenagers with DS and congenital cardiopathy

Variables	n (%)
Patients	98 (100)
Females	55 (56,1)
Males	43 (43,9)
Age	
0 to 2 years	24 (25)
2 to 6 years	23 (23)
6 to 10 years	11 (11)
10 to 18 years	40 (41)
Gestational Age	
Term	76 (77,6)
Pre-term	22 (22,4)
Maternal Age	
Up to 20 years	4 (4)
21 to 34 years	29 (30)
More than 35 years	65 (66)
Maternal Schooling	
No answer	2 (2)
Illiterate	0 (0)
Primary School (Incomplete)	43 (43,9)
Primary School (Complete)	17 (17,3)
High School (Incomplete)	5 (5,1)
High School (Complete)	24 (24,5)
Higher Education (Incomplete)	3 (3,1)
Higher Education (Complete)	4 (4,1)
Cardiac Surgical Procedure	49 (50)
Up to 2 years	38 (39)
2 to 6 years	8 (8)
More than 6 years	3 (3)
Type of Cardiopathy	
AVSD	49 (50)
IVC	27 (28)
IAC	23 (24)
PAD	21 (22)
TOF	13 (13)

AVSD – Atrioventricular Septal Defect; IVC - Interventricular Communication;
IAC - Interatrial Communication; PAD – Patent Arterial Duct; TOF – Tetralogy of Fallot.

for their ages (table 2, figure 1 and 2).

There was a statistically significant difference

between malnutrition frequency at birth and current nutrition status of the studied population based on

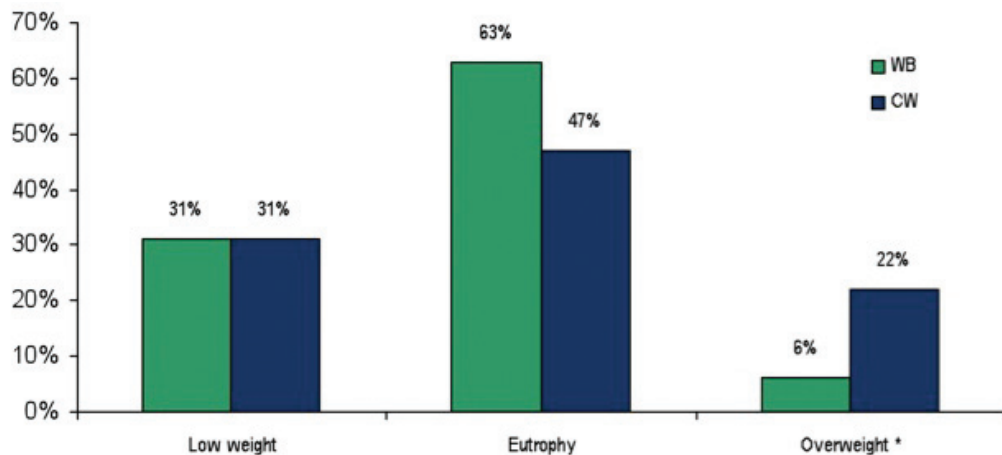
TABLE 2

Birth and current anthropometric data of the 98 children and teenagers with DS and congenital cardiopathy.

Variables	n (%)
Patients	98 (100)
Weight at Birth	
Low weight	30 (31)
Eutrophy	62 (63,3)
Overweight	6 (6,1)
Stature at Birth	
Low Stature	41 (42)
Regular Stature	57 (58,2)
Difficulty to Gain Weight	51 (52)
Adequate Gain of Weight	26 (26)
Current Weight	
Low weight	30 (31)
Eutrophy	46 (46,9)
Overweight	22 (22)
Current Stature	
Low Stature	17 (17)
Regular Staturel	81 (82,7)

FIGURE 1

Comparison between birth and current W/A.



* p= 0,049; WB: Weight at birth; CW: Current weight.

anthropometric data (52% Vs 34%) ($p<0.01$) (table 3). The relationship between age and malnutrition showed a border line linear association ($p=0.057$) of nutritional status improvement as age increased.

The connection between current and birth W/A ratio showed significant worsening in the nutritional status linked to overweight ($p<0.05$). Furthermore, there was statistical significant association ($p<0.001$) between birth and current S/A, pointing towards a stature growth recovery (figures 1, 2).

The most prevalent maternal education status, when added to birth and current nutritional status, was the primary school level (completed or not), although not significantly associated to malnutrition.

The study did not show significant results when

relating cardiopathy and malnutrition, nor was any nutritional status improvement noticeable after cardiac surgical procedure.

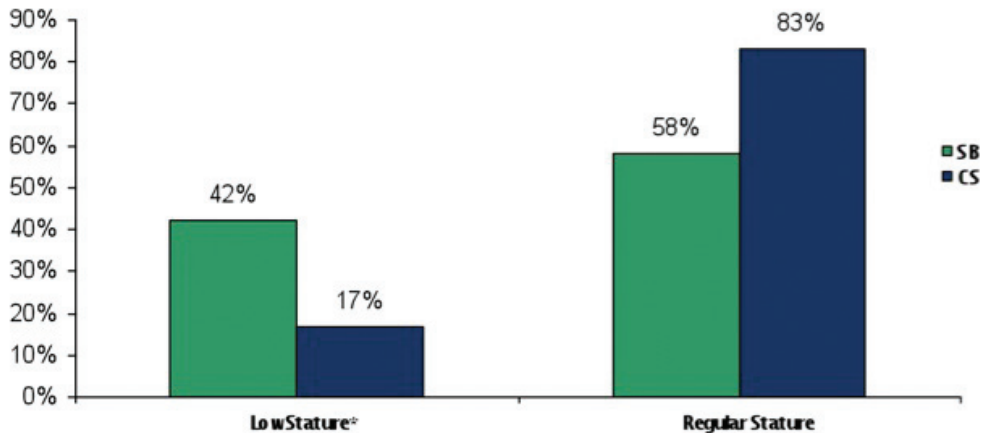
DISCUSSION

This cross-sectional study on children and teenagers with DS and congenital cardiopathy showed that such patients present stature and weight deficit at birth. As age increases, there is stature growth improvement, however this is not properly followed by weight. At contrary, it was observed that these patients show a greater propensity towards overweight along the years.

The Moura and collaborators study¹⁸ which evaluated 18 individuals, being 6 males (33.3%) and 12 females (66.6%), showed similar results to ours, presenting

FIGURA 2

Comparison between birth and current S/A.



* $p=0.049$; SB: Stature at birth; CS: current stature.

TABLE 3

Relation between birth and current malnutrition of the 98 children and teenagers with DS and congenital cardiopathy.

Variables n (%)

Birth malnutrition	51 (52)
Current malnutrition	33 (34)

* $p=0.01$

greater prevalence of DS and congenital cardiopathy in females.

As for prematurity, it was not found any association between premature birth and DS or congenital cardiopathy. In our study, as well as in Theodoro's (19), there was greater prevalence of term births.

According to Freeman and collaborators (20), more studies on the relation of maternal age and DS are necessary, although advanced maternal age is a risk factor for DS. At the biological point of view, what is more important is the physiological age of the ovary. One possible explanation would be that at advanced age there is ovule depletion. Then, women with reduced number of ovules, for other reasons, may have greater risk of conception with a trisomy of chromosome 21. The Binkert and collaborators (21) study, for instance, indicated positive correlation between the mothers' advanced age and the risk of conceiving a DS child, although the pathological mechanism remained unclear.

In line with the study of Lopes and collaborators (15), as well as Zini (22), regarding maternal education, most mothers had not completed primary school.

The individuals of the study showed impairment in weight and stature development, being both significantly lower than normal, in agreement with the results of other studies (10-12, 15, 23). Theodoro (19) highlights that babies with DS are born with discrete retardation in their growth and with lower percentiles regarding the ratio weight/stature. This particular growth profile is characterized by a premature trigger of the growth spurt and reduced linear growth velocity, which result in individuals with lower stature than the general population (7,8,12). Besides that, there is propensity for overweight, usually starting from puberty. This fact is related to growth deficit itself, which determines a lower basal metabolic rate (7,14,24-26). According to Moura and collaborators (18) and Benato and collaborators (27), DS individuals present greater probability of thyroid dysfunction (hypothyroidism), which can be the cause for overweight and obesity. These studies suggest that the slower basal metabolic rate, the compulsive feeding due to difficulty in chewing and the general hypotony of the muscles (including those involved in digestion) as possible causes of obesity in these individuals. Specially due to the hypotony of the muscles involved in digestion, DS patients experience less satiation feeling after a meal, and this make them eat more.

Theodoro (19) considers the low stature a contributing factor to obesity, since this aspect diminishes the daily energetic need and consequently contributes for the caloric ingestion above necessary. He also links obesity with chronic diseases in adulthood, especially cardiac

ones. Therefore, children with DS who develop obesity in their first year and remain obese until adulthood have greater risks of developing cardiovascular diseases. Silva e collaborators (28) report a greater concentration of visceral fat and higher percentage of overweight in people with DS If compared to regular population.

Benato and collaborators (27) show that children with DS in association with severe congenital cardiac disease present feeding problems and weight gain below expected. Many aspects of the treatment of such cardiac diseases may affect the ability of these children to increase weight normally, among them: admissions, treatment and/or surgery, all leading to difficulties in weight gain and resulting in a low weight child (29). Respiratory and suction problems are also clinical manifestations that commonly lead to nutritional difficulty in this group. In cardiopathic infants, the act of sucking is an activity that requires a lot of effort. It can cause excessive fatigue and aggravate the dyspnea scenario. In older children, the respiratory difficulty and fatigue interfere with the capacity of chewing and swallowing (30). However, as soon as the congenital cardiac defect is repaired, children with DS start to increase weight adequately. We believe that the absence of statistical significance in the association between cardiac defect repair and nutritional status improvement observed in this study may be explained by the small size of the analyzed sample and/or the lack of a control group composed of patients who do not have congenital cardiopathy in our study.

Others have found a connection between the frequency of malnutrition at birth and later on in life. These studies present evidence suggesting that as age increase, the nutritional status improve. Nevertheless, if on one hand there has been stature growth recovery based on S/A ratio data, on the other hand there has been tendency towards overweight from W/A ratio data (7,14,24-26).

In a study by Cronk (12) made in England with 262 DS patients between 2 and 18 years of age, the authors showed that at least 30% of the individuals presented some degree of overweight (22).

Gomes and collaborators (31), in a study designed to investigate the incidence of overweight and obesity in children with DS, it has been found that 83% of children with ages between 2 and 6 are within the normal weight standards, while 17% are overweight or obese (31,32). In other two studies, by Giaretta and collaborators (4) and Fernahall (33), it was noticed that 33.3% of adolescents with DS are overweight, 33.3% are well-nourished and 33.3% malnourished. These findings reinforce the results found in other studies on children and teenagers with DS – which showed an increase in the prevalence of obesity ever since the childhood (32,33). This data

demonstrates the importance of the nutritional follow-up in order to prevent overweight and obesity of this population (32,33).

As in the work by Ferrin and collaborators (34), our study showed prevalence in the type of congenital cardiopathy found – half of the evaluated individuals presented AVSD (8-11). However, there was no association between the type of cardiopathy and malnutrition, nor there was any nutritional status improvement after cardiac surgical procedure.

The information found in studies such as this one is of great value for establishing a general profile of patients with DS and congenital cardiopathy. Such information is also important as basis to new researches, since this is an understudied population. Furthermore, DS populations have gone through significant profile changes over the past years, especially due to the increase in their life expectancy.

The existing curves used in this study do not show a specific cut-off point for the population with DS; thus, it was chosen to use the standard cut-off points recommended by the international organizations for children and teenagers without DS. It was noticed that DS children, just like the regular population, also need to be anthropometrically evaluated based on curves of W/S and Body Mass Index for age (BMI/A), since the existing curves underestimate excess of weight and overestimate low weight of these individuals.

CONCLUSION

The individuals evaluated in this study presented stature and weight deficit at birth and, as age increased, they improved their stature profile. We can no longer link DS and congenital cardiopathy to malnutrition. Nowadays, these patients have been going through a nutritional transition: they are becoming individuals with overweight and obesity and leaving the malnourished group they used to be part of.

It is during puberty that these patients are developing overweight, and that is reflecting a greater risk for the development of chronic diseases in adulthood. Our study showed evidence of high overweight prevalence among those people. Therefore, public health care policies should take into consideration the particularities of the DS population. In addition, new international growth curves must be constructed and consider the comorbidities associated with this population.

RESUMEN

El objetivo de este estudio fue analizar el perfil nutricional de las pacientes con síndrome de Down (SD) y cardiopatía congénita (CC). Se trata de un estudio

transversal de evaluación del estado nutricional en 98 pacientes con SD y CC, 56% mujeres entre 0 y 18 años de edad. La prevalencia de bajo peso, sobrepeso y baja estatura fue de 30%, 22% y 17%, respectivamente. Hubo diferencia entre la desnutrición al nacer y la que se refiere a los actuales datos antropométricos ($p = 0,01$). Una asociación mostró un incremento significativo en el peso ($p = 0,049$) y hubo recuperación de la estatura ($p < 0,001$). En conclusión, el nacimiento hay déficit de estatura y peso. Al aumentar la edad hay una mejoría en la estatura, pero no así en el peso. Se observó una predisposición al sobrepeso en estos pacientes.

Palabras clave: síndrome de Down, cardiopatía congénita, evaluación nutricional, obesidad, malnutrición.

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