On the Gravity of the Acute Rheumatic Fever in Children from Pernambuco, Brazil

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Introduction

In an anatomopathological study dated 1970, Lira et al[1] showed the severity of Acute Rheumatic Disease (ARD) in Pernambuco, describing 43% of the 52 cases studied in childhood and highlighting the importance of cardiomegaly and the high level of adhesive pericarditis. This study contradicted the idea that the ARD was a condition inherent in cold climates. Although in almost all regions of the world the reduced incidence and increased prevalence of the disease vis-à-vis the application of Doppler echocardiogram are described in the study of populations[2], in our field, despite parallels with this universal finding, severe forms of ARD arise, requiring early surgical management of heart valve lesions in children with high surgical risk.

Why Pernambuco still presents such severe forms of a disease nearly extinct in developed countries? The analysis of the clinical condition of 13 severely ill children, studied in detail for a short period - 18 months - at a single hospital in Recife, out of 54 children, thus revealing high prevalence, should partly answer this question.

Clinical characteristics of a sample of 13 patients

From January 2011 to June 2012, 54 children with acute rheumatic heart disease, with diagnosis based on the modified Jones criteria, assisted at the IMIP, 13 of which were hospitalized with a severe clinical picture, accounting for 24.2% of this series, with active ARD. In a recent hospital study conducted in Auckland, New Zealand[3], over a 12-year period, 44 patients were described, which shows the representativeness of the sample, obtained in a short period of 18 months.

Table 1 shows clinical and laboratory data that caught our attention. In the analysis, we can see that:

a) The picture of rheumatic heart disease was preceded by tonsillitis in at least half of the cases, with fever and arthritis in nearly 70% of them;

b) Congestive Heart Failure (CHF), including Acute Pulmonary Edema (APE), occurred in 100% of patients with Mitral Insufficiency (MI) diagnosed in the same 100%, accompanied by Aortic Regurgitation (AR) in about one third of the cases – in the genesis of IM, the rupture of the mitral valve chordae tendineae in ¼ of the series was relevant. Despite the CHF, the left ventricular ejection fraction (LVEF) remained normal or exaggerated, except in 2 patients with MI with ruptured chordae tendineae and AR – normal LVEF is consistent with the literature, a fact that comes in disfavor of a "myocardial factor" in the genesis of the CHF, which would be due primarily to the valvular involvement[4];

c) Only one case of chorea (case 2 - 7.7%) was observed;

d) There was severe cardiomegaly with average cardiothoracic index (CTI) of 57.7%, reaching as much as 71.7%;

e) On three occasions, very high values were found for the number of leukocytes in peripheral blood, and, on four occasions, there were high levels of Anti-streptolysin O (ASO), contradicting what is put by Décourt[5], who recognizes a slight increase in these variables, arguing marked bacterial aggressiveness and long-lasting antigenic stimulation;

f) The QTc value - a potential indicator of severity in ARD[5] - proved to be increased in three patients (cases 4, 6 and 8 - 23.0%), according to Décourt values. On the ECG, we saw in a patient with "extreme generalized edema" (!) – an old condition described by Bouillaud, in France, in 1836, in a 30-year-old man[6] - "fragmented QRS complex", in extrasystoles originating from the right ventricle, suggestive of the possibility of sudden death[7], as well as the presence of "inverted U waves" in the left precordial leads, emerged shortly, indicating severity of ventricular overload, almost always present in sick patients (first-degree AV block was seen on two occasions - 15.3%);

g) Of the 13 patients, 10 (76.9%) underwent implantation of bioprosthetic valves in valves mutilated by rheumatism.

Nutritional and socioeconomic characteristics of the sample

Table 2 presents the most significant findings.

Thus,

a) With a mean age of 8.5 ± 3.2 years, and a slight male predominance, they came from all regions of the state of Pernambuco. This average age is below the number that is considered most frequent: 10 years[8];

b) Crowding, one of the main factors in the genesis of ARD, was far from the high values found in Australia[9] - 6.9/7.5 persons

Keywords

Rheumatic Fever; Rheumatic Heart Disease; Heart Failure; Heart Valve Diseases; Social Conditions; Poverty.

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Table 1 – Clinical and laboratory findings in 13 children with severe acute rheumatic fever. Recife, 2013

Case	Fever	Tonsillitis	Arthritis	Carditis	CTI (%)	ASO (UI)	Leukocytes /mm^3	Hb (g/dL)	ESR (mm)	QTc (s)	Doppler echocardiography	
											LVEF (%)	Valvular lesions
1	S	S	N	CHF – Chest Pain	71.7	2.240	14.100	11.6	40	0.400	60	MR — Rupture of chordae tendineae — Pericarditis
2	N	N	N	Dyspnea	55.5	-	-	-	-	0.413	53	MR + AR
3	N	N	S	CHF	57.7	419	11.000	10.5	28	0.352	72	MR
4	N	N	S	Generalized edema	56.7	297	13.200	9.6	50	0.434	67	MR + AR
5	S	N	S	CHF	65.2	1.130	13.000	10	100	0.405	55	MR — Rupture of chordae tendineae + AR
6	S	N	N	CHF	55.0	212	14.700	10.6	72	0.425	76	MR
7	S	S	S	Chest pain - Palpitations	50.0	-	13.200	12.3	19	0.351	67	MR
8	S	S	S	CHF	66.7	1.091	12.200	9.9	45	0.447	60	MR + AR
9	S	S	S	CHF	54.7	200	7.200	11.7	22	0.407	69	MR – Rupture of chordae tendineae
10	N	N	S	APE	61.0	170	27.000	11.4	65	-	62	MR + AR
11	S	S	S	CHF	53.0	220	8.200	9.7	72	0.398	70	MR
12	S	S	N	CHF	47.5	218	7.200	12.2	20	0.400	65	MR
13	S	N	S	CHF – Pneumonitis	55.5	4.030	17.200	13.5	35	0.388	65	MR
% and Mean +Standard deviation	69.2%	46.2%	69.2%	100%	57.7 ± 6.9			11.1 ± 1.2	47.3 ± 25.4	0.402 ± 0.03	64.7 ± 6.6	MR – 100% AR – 30,8% Rupture of chordae tendineae – 23.1%
Median						297	13.100					

CTI: Cardiothoracic index; ASO: Anti-streptolysin O levels in peripheral blood; HB: Hemoglobin; ESR: Erythrocyte sedimentation rate; LVEF: Left ventricular ejection fraction; CHF: Congestive heart failure; MR: Mitral Regurgitation; AR: Aortic regurgitation.

per bedroom — resulting in an average of 1.5 person/room, which is little significant. But we had 2 patients living in the slums of Recife (cases 9 and 11), residing in "houses" made of wood/cardboard in a "small interspace", which includes a living room, a bedroom and a kitchen;

c) The per capita income revealed underprivileged families (average of R$ 138.20), two of which in a condition of poverty, according to the federal government (cases 2 and 4);

d) The nutritional study showed that the patients had adequate height, but the analysis of body mass index (BMI) revealed that 6 of them were "skinny" (BMI -2 z scores), in a severe way (case 10, BMI -3 z scores) due to recent weight loss. For this purpose, in Pernambuco, recent research on the behavior of the height of our children found that there was an increase in this variable from 1945 to 2006, revealing better feeding conditions in the early years of life, since the height is the most faithful ecological parameter that allows the genotypic growth factor to express freely in this stage of life[10].

Characteristics peculiar to patients coming from rural areas

Three patients coming from rural areas (4, 8 and 10) lived in modest isolated homes, in small communities far from urban areas, failing to recognize basic clinical symptoms, such as "sore throat": since the family members are unaware of rheumatic fever, children suddenly present dramatic expressions of clinical conditions, contrasting with the epidemiological factors of the disease, especially the events necessary for the emergence of streptococcal strains[5,8], which requires the application of aerosols of the micro-organism "from mouth to mouth," under a situation of overcrowding. Late diagnosis of streptococcus probably induces the bacteria to constantly stimulate the immune system.

Conclusions

In Pernambuco, there are still severe forms of ARD, similar to those described by Lira et al[1] 42 years ago. However, we cannot speak of "poverty clusters," since they come from all regions of

Table 2 – Socioeconomic and nutritional aspects in 13 children with severe rheumatic disease. Recife, 2013

Case	Age (y)	Sex	Origin	Family composition	Rooms per home	Per capita Income (R$)	BMI (z scores)	H/A (z scores)
1	4.2	F	Rural Area	3	3	414.00	-2	1
2	10.11	F	Capital	7	2	77.14	MD	MD
3	7.6	M	RMR	5	4	143.60	-2	1
4	13	F	Rural Area	6	4	34.00	MD	-1
5	6	M	Wild rural area	4	4	260,00	MD	-1
6	4.3	M	Rural area	4	3	-	1	-1
7	6.4	M	Capital	4	4	286,25	-1	MD
8	6	M	Rural area	2	2	130,00	-2	1
9	9	M	Recife	2	1	311.00	-2	MD
10	13.7	M	Rural area	4	2	400.00	-3	MD
11	12	F	Capital	6	1	132.70	1	MD
12	11	F	Forest	5	5	81.60	MD	-1
13	7.6	M	Wild rural area	7	4	86.00	-2	2
Mean ± Standard deviation	8.5 ± 3.2			4.5 ± 1.7	3 ± 1.3			
Median						138.2		

Y = Years; BMI = Body mass index; H/A = Nutritional indicator height/age (WHO); MRR - Metropolitan region of Recife.

the state. Poverty, low per capita income, poor housing[8], and especially lack of diagnosis of streptococcal pharyngitis are the factors involved. The main clinical expression is carditis with CHF, which include mitral regurgitation with ruptured chordae tendineae, cardiomegaly, adhesive pericarditis, pneumonitis and unusual electrocardiographic aspects. Therefore, the state of Pernambuco presents an unfavorable social situation that induces the appearance of aggressive streptococcus strains, perhaps rich in M protein[5]. Primary prevention would result from better housing and hygiene[8].

In our group, Santos calls attention to an intriguing fact: the disproportion between the small number of children diagnosed with ARD and the large number of adults with rheumatic valve disease, accounting for 40% of cardiac surgeries in Brazil, as if between these two extremes, a large number of patients did not have the disease recognized: would subclinical carditis be relevant here? Therefore, cases diagnosed between 5 and 14 years could be included in the expression "the tip of an iceberg" only.

Author contributions

Conception and design of the research: Saraiva LR; Acquisition of data: Ventura C, Sobral MA, Barbosa B, Moraes F; Analysis and interpretation of the data: Saraiva LR, Santos CL; Statistical analysis: Parente GB; Critical revision of the manuscript for intellectual content: Santos CL; Realization of surgery: Moraes F

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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