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. In a recent hospital study conducted in Auckland, New Zealand\(^1\), 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\(^2\) - 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, 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.

Keywords

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

Mailing address: Lurildo Ribeiro Saraiva • Estrada do Arraial, 2405704, Tamarineira. Postal Code 52051-380, Recife, PE - Brazil
E-mail: lurildo@cardiol.br, lurildocleano@hotmail.com
Manuscript received January 11, 2013; revised manuscript January 15, 2013; manuscript accepted April 23, 2013.

DOI: 10.5935/abc.20130172
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/mm³ | Hb (g/dL) | ESR (mm) | QTc (s) | Doppler echocardiography | LVEF (%) | Valvular lesions |
|------|-------|-------------|-----------|----------|---------|----------|----------------|-----------|----------|--------|--------------------------------|---------|-----------------|
| 1    | S     | S           | N         | CHF      | 71.7    | 2.240    | 14.100         | 11.6      | 40       | 0.400  | MR — Rupture of chordae tendineae — Pericarditis | 60      | MR + AR          |
| 2    | N     | N           | N         | Dyspnea  | 55.5    | -        | -              | -         | -        | 0.413  | MR + AR                          |
| 3    | N     | N           | S         | CHF      | 57.7    | 419      | 11.000         | 10.5      | 28       | 0.352  | MR                              |
| 4    | N     | N           | S         | Generalized edema | 56.7  | 297      | 13.200         | 9.6       | 50       | 0.434  | MR + AR                          |
| 5    | S     | N           | S         | CHF      | 65.2    | 1.130    | 13.000         | 10        | 100      | 0.405  | MR — Rupture of chordae tendineae + AR | 55      | MR + AR          |
| 6    | S     | N           | N         | CHF      | 55.0    | 212      | 14.700         | 10.6      | 72       | 0.425  | MR                              |
| 7    | S     | S           | S         | Chest pain - Palpitations | 50.0  | -        | 13.200         | 12.3      | 19       | 0.351  | MR                              |
| 8    | S     | S           | S         | CHF      | 66.7    | 1.091    | 12.200         | 9.9       | 45       | 0.447  | MR + AR                          |
| 9    | S     | S           | S         | CHF      | 54.7    | 200      | 7.200          | 11.7      | 22       | 0.407  | MR — Rupture of chordae tendineae | 69      | MR + AR          |
| 10   | N     | N           | S         | APE      | 61.0    | 170      | 27.000         | 11.4      | 65       | -      | MR + AR                          |
| 11   | S     | S           | S         | CHF      | 53.0    | 220      | 8.200          | 9.7       | 72       | 0.398  | MR                              |
| 12   | S     | S           | N         | CHF      | 47.5    | 218      | 7.200          | 12.2      | 20       | 0.400  | MR                              |
| 13   | S     | N           | S         | CHF      | 55.5    | 4.030    | 17.200         | 13.5      | 35       | 0.388  | 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 ± 0.402 ± 0.03 ± 64.7 ± 6.6

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.

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 strains5,8, 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.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, and especially lack of diagnosis of streptococcal pharyngitis are the factors involved. The main clinical expression is carditis with CHF, which includes 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. Primary prevention would result from better housing and hygiene.

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.

Sources of Funding
There were no external funding sources for this study.

Study Association
This study is not associated with any post-graduation program.
References

1. Lira V, Freitas D, Maciel SM. Estudo morfológico da cardiopatia reumatismal em Recife (Brasil). An Fac Med Univ Fed Pe. 1970;30:145-62.

2. Marijon E, Ou P, Cefermazier DS, Ferreira B, Mocumbi AO, Jani D, et al. Prevalence of rheumatic heart disease detected by echocardiographic screening. N Engl J Med. 2007;357(5):470-6.

3. Siriett V, Crengle S, Lennon D, Stonehouse M, Cramp G. The epidemiology of rheumatic fever in the Tairawhiti/Gisborne region of New Zealand: 1997-2009. N Z Med J. 2012;125(1365):8-15.

4. Veasy LG, Tani LY. A new look at acute rheumatic mitral regurgitation. Cardiol Young. 2005;15(6):568-77.

5. Décourt LV. Doença reumática. 2ª ed. São Paulo: Savier; 1972.

6. Bouillaud J. Nouvelles recherches sur le rhumatisme articulaire aigu en général. Paris: Chez J - B. Baillière; 1836. p 50.

7. Pei J, Li N, Gao Y, Wang Z, Li X, Zhang Y, et al. The J wave and fragmented QRS complexes in inferior leads associated with sudden cardiac death in patients with chronic cardiac failure. Europace. 2012;14(8):1180-7.

8. Carapotis JR, Steer AC, Mulhalland EK, Weber M. The global burden of group A streptococcal diseases. Lancet Infect Dis. 2005;5(11):685-94.

9. Brown A, McDonald MI, Calma T. Rheumatic fever and social justice. Med J Austr. 2007;186(11):557-8.

10. Figueiroa JN, Alves JG, Lira PI, Batista Filho M. Evolução intergeracional da estatura no Estado de Pernambuco, entre 1945 e 2006. 2- aspectos analíticos. Cad Saúde Pública. 2012;28(8):1468-78.