NASCER E CRESCER
BIRTH AND GROWTH MEDICAL JOURNAL
year 2019, vol XXVIII, n.º 4

Nascer e Crescer - Birth and Growth Medical Journal
2019;28(4): 174-178. doi:10.25753/BirthGrowthMJ.v28.i4.19085

EDITORIAL

PEDIATRIC CARDIOLOGY IN THE 21st CENTURY: ADVANCES AND CHALLENGES

A CARDIOLOGIA PEDIÁTRICA NO SÉCULO 21: AVANÇOS E DESAFIOS

Cleonice C. Mota[I]

In the 21st century, technological progress in diagnosis and treatment of cardiac diseases is getting more intense and extensive. In paediatric cardiology, a repetition of what has been observed in other bioscience areas is evident: a disordered technological advance involving little critical analysis. This fast pace in the technological incorporation raises ethical questions — should we do everything we possibly can? Political and economic questions should also be asked — how, when, how much, and who is responsible for allocating the available resources?

This issue acquires ethical and political dimensions due to both the rapidly growing gap resulting from the increasing demand and the limitation of resources — a real problem, even in the wealthiest economies in the world, and a scandalous drama in the poorest ones. There is no solution in sight but it is a challenge that cries out for urgent discussion.

In low- and middle-income countries, estimates show that 15 million children become disable or die every year from heart disease and the treatment is inaccessible or of low-quality for 90% of this population.[1]

Through technological progress, new clinical and surgical therapeutic options have been incorporated into the routine practice of paediatric cardiology. Complex congenital heart diseases previously impossible to be addressed are now satisfactorily managed with surgical approach. Some syndromes such as trisomy 13 also called Patau syndrome — characterized by brain malformations, facial dysmorphism, eye abnormalities, polydactyly, severe psychomotor delay, and visceral malformations — is associated with cardiac malformations in 80% of cases. Trisomy 18, also known as Edward's syndrome, is associated with a very high rate of multiple congenital heart defects. Despite the severity and the high risk of fetal and neonatal death, long survivals have been reported in children presenting these two syndromes: 27 years for trisomy 13 and 50 years for trisomy18.[2,3] This poses new and difficult challenges for the paediatric cardiologist. How to manage patients with cardiac diseases and these syndromes which are considered lethal but not always incompatible with life and may present survivals of months, years or even decades? This is accomplished through an increasingly multidisciplinary approach, with the family playing a central role in the decision process.

When handling complex situations such as these, dealing with ethical, legal, religious, and cultural aspects in populations with increasingly diverse values and beliefs, it calls for the healthcare professionals to acquire new communication skills and knowledge of values and beliefs different from their own.

Intrauterine surgery in fetuses with vascular and cardiac malformations, previously unthinkable to be treated, became possible in Fetal Medicine Units. Invasive treatment of congenital and acquired heart defects, previously only approached through surgery, can now be alternatively addressed through cardiac catheterization. Advanced intensive life support keeps newborns and children with extremely complex cardiac diseases alive and with chances of recovery.

I. Professor at Faculty of Medicine, Universidade Federal de Minas Gerais. 30130-100 Belo Horizonte-MG, Brazil
cleomota@medicina.ufmg.br

New drugs with greater therapeutic potential, such as monoclonal antibodies and new antihypertensive agents have been used, but with much higher costs than those routinely employed. Some of these new drugs cost many thousand dollars or euros per month and are chimeras for the overwhelming majority of those who need them.

In this context, paediatric cardiology then faces a double challenge. Just like Janus, the Two-Faced God, it must look forward to a technologically promising future — achievable? — but also bearing in mind the past, which persists. Despite all advances, we have to deal with the persistence of neglected diseases in low-resource regions and their reappearance in high-income locations thus posing a problem for many of the young cardiologists who have never handled them.

The case of rheumatic fever and its sequel, the rheumatic heart disease, is emblematic because it poses a double challenge. Our prospects include more refined surgeries for the treatment of valvular lesions, expansion of existing services, health care in-person and at distance, permanent education for health professionals and the very real prospect of a vaccine against the streptococcal infection. On the other hand, in 2010, it was estimated a worldwide prevalence of 34.2 million individuals with rheumatic heart disease, resulting in 345,110 thousand deaths and the loss of 10.1 million DALYS —disability-adjusted life years — every year.[4-6] This epidemiological data impel the need to look back to the past. Unlike regions where the disease has already been eradicated after the steps taken in its control, we are faced with its persistence in marginalized communities without effective programmes of prevention. Nowadays, we still have to deal with the scandal of the lack of such basic, inexpensive, and easy to administer input as benzathine penicillin, exclusively and obscenely driven by market considerations and public health policy issues — or their lack — , instead of focusing the best interest of populations, who mostly need care.

Despite the knowledge acquired in recent decades, rheumatic heart disease remains the most prevalent acquired heart disease in individuals under the age of 25.[7] The potential implications affect all age groups, considering that valve sequels are present throughout life, and also determine premature deaths. Recently, a large-scale study reported 16.9% of deaths in the considered population, with a mean age of 28.7 years.[8] In addition to the high social costs and mortality rates in the poorest population, the disease is at risk of re-emerging in areas where it had been previously controlled.

In 2017, R$157,578,000.00 (around 40 million dollars) were spent by the Brazilian Public Health System (SUS) on hospitalizations due to rheumatic fever and rheumatic heart disease. Out of all cardiac surgeries at that time, 31% were performed on patients with rheumatic valvular lesions.[9] As regards the economic impact of primary and tertiary preventions, it has been shown a marginal cost of US$46 for diagnosis and treatment of a streptococcal pharyngotonsillitis with one benzathine penicillin injection and the total annual cost of US$319 per patient for the therapeutic approach of rheumatic fever and its sequels.[10,11] This analysis demonstrates the need to eliminate barriers for the disease control. In this context, it should be considered the high incidence of rheumatic fever and high prevalence of rheumatic heart disease, in addition to the increasing severity of valvular lesions with recurrences.

As for prospects to modify the current scenario, the disease has received increasing attention in recent years through regional initiatives focused on prevention programmes and with the support of several national and international organizations such as WHO, WHF, and AHA. Additionally, acquisition of comprehensive and high-quality data is crucial for the development of strategies to control the disease. Among the priorities to address contemporary data, major efforts have been directed towards investigations in high prevalence sentinel areas and also carried out in different geographical regions.[12] Contributions of Working Groups in strategic planning of access to diagnosis, treatment and prevention, particularly the setting up of the "WHF Taskforce on Rheumatic Heart Disease", are also initiatives aligned with the purpose of modifying the epidemiological profile of the disease and provide sustainability for the projects. Both actions are in agreement with the "Global Resolution on Rheumatic Fever and Rheumatic Heart Disease" of WHO approved in 2017, when the disease was recognized as a global health priority.

Despite many unanswered questions and the magnitude of the challenge, concerted efforts have to be made to seek data through scientific investigation and to implement effective health policies. Allocation of resources and access to diagnosis and treatment as well as comprehensive prophylaxis programmes, are key to minimize the burden of rheumatic fever and the onus to be paid by its sequels.

The example of rheumatic fever is reproduced in several other areas of paediatric cardiology, in which the unequal and unfair distribution of financial and technological resources generate islands of excellence in the provision of health services surrounded by an ocean of unassisted children and adolescents deprived of high-quality care.

Addressing and dealing with this dilemma with both technical and political actions is the most urgent challenge for us, paediatric cardiologists in the 21st century.

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Neste século XXI, o incremento tecnológico na área da saúde é cada vez mais intenso e extenso. Na cardiologia pediátrica repete-se o que ocorre em outras áreas da biociência, um avanço tecnológico desordenado e pouco crítico. Esta velocidade de incorporação tecnológica levanta questões éticas – devemos fazer tudo que podemos? E questões político-econômicas – como, quando, quanto e a quem cabe a responsabilidade de alocar os recursos disponíveis?

Pela limitação dos recursos – problema real, mesmo nas economias mais ricas do mundo e drama escandaloso nas regiões mais carentes – que enfrentam uma defasagem rapidamente crescente com as demandas das pessoas na área da saúde, a questão assume um perfil ético e político, sem solução à vista, mas que clama por discussão urgente.

Nos países de baixa e média renda, estima-se que, anualmente, 15 milhões de crianças tornam-se doentes ou morrem em decorrência de doenças cardíacas e o tratamento é inacessível ou de baixa qualidade para 90% dessa população.[1]

Pelo avanço tecnológico, novas possibilidades de tratamento, clínico e cirúrgico, são incorporadas à rotina da cardiologia pediátrica. Algumas síndromes, antes consideradas inabordáveis do ponto de vista cardiológico, hoje são algumas vezes satisfatoriamente conduzidas com tratamento cirúrgico. A trissomia do 13 ou síndrome de Patau – caracterizada por malformações cerebrais (holoprosencefalia), dismorfismo facial, anomalias oculares, polidactilia, atraso psicomotor grave e malformações viscerais –, é associada com malformações cardíacas em 80% dos casos. A trissomia do 18 ou síndrome de Edwards também apresenta um índice muito elevado de lesões cardíacas múltiplas. Apesar da gravidade e do alto risco de morte fetal ou no período neonatal, há relatos de longas sobrevidas por décadas em crianças com essas duas síndromes: no caso da trissomia do 13 por 27 anos e no caso da trissomia do 18 por 50 anos.[2,3] Isso gera novos e difíceis desafios para o cardiologista pediátrico: como lidar com um diagnóstico de malformação dita letal, e que nem sempre é incompatível com a vida e, às vezes, apresenta sobrevidas de meses, anos e até décadas? Essa abordagem é cada vez mais multiprofissional, com a família tendo um papel central nas decisões.

Na perspectiva de situações complexas como essas, lidar com aspectos morais, éticos, legais, religiosos e culturais em populações cada vez mais diversas em valores e crenças, implica a necessidade do profissional de saúde adquirir novas habilidades comunicativas e conhecimentos de valores e crenças diversas das suas.

Cirurgias intraútero em fetos com defeitos vasculares e cardíacos, anteriormente sem possibilidade de tratamento, tornam-se rotineiras em serviços de medicina fetal. Tratamentos invasivos dos defeitos cardíacos congênitos e adquiridos, antes realizados somente devido ao avanço das técnicas cirúrgicas, hoje tem alternativa de intervenção