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Cystic fibrosis in Brazil: the pediatrician's turn

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Lessons learnt: undeniable advances, multiple and complex challenges. Also multiple are the teachings of the group from Campinas contained in their article published in this issue of the *Jornal de Pediatria*; they toast us with a wide-ranging, competent and provocative report on their sample of 104 cystic fibrosis patients.¹ Most probably the richest work in specialized Latin-American literature.

For these reasons, the aspects to be analyzed are equally multiple. An agreeable, although to a certain extent ungrateful, task, since editorial space, by nature, is limited. Limitation does not conjugate well with multiplicity and so we are obliged to isolate those aspects that will be analyzed with a view to the general pediatrician and those that work in related subspecialties (pulmonology, gastroenterology, nutrology and others), also pediatricians first and foremost. With space at a premium, we, intentionally, choose two items: survival and age at diagnosis because both reflect on the (lack of) care that patients have been receiving, and which is not limited to this, esteemed, center of excellence.

See related article
on page 371

In survival, we have made progress. Brazilian CF centers have participated in two collaborative studies,^{2,3} that covered the periods 1979-1989 and 1960-1989 and onto which were enrolled respectively, 743 and 1,827 patients, from four and ten Latin-American countries. Mean survival age at death was just 6-7 years. Brazilian centers integrated the both studies and the mean age of patients who were still in follow-up from the first period (1979-1989) was just 6.4 years.² Another Brazilian study covered a period starting before and ending after these two studies, and performed at a service that also took part in the two previously mentioned studies, found that for the period 1970-1994, mean survival had jumped to 12.6 years.⁴ Another pleasant surprise is afforded us by the cohort from Campinas: during the decade 1990-2000, median survival reached a new level of 18.4 years of survival age after diagnosis,¹ an estimate which is equivalent to figures from the United States in the 80s. As the authors explain, life expectancy with this condition in industrialized nations is currently around 31.6 years.¹ Therefore, we could ask: would our general pediatricians win our patients extra years, through greater proximity and more frequent consultations, if there was a directed effort in combination with the sub-specialists?

With respect of age at diagnosis, we have remained parked. For the two Latin American cohorts and the cohort evaluated by the Campinas team, average age at diagnosis was, respectively, greater than 3-4 years^{2,3} and 4 years and 2 months.¹ Notwithstanding, it is enough to observe in the article that is the subject of this Editorial that 81.1% of the

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patients were already symptomatic during the first year of life, a finding which is comparable with a series of cases studied by a group from Ribeirão Preto, São Paulo.^{1,5} Once more, we could ask: would patients be diagnosed earlier if sub-specialists could count on the cooperation of general pediatricians?

The non-existence of a partnership with pediatricians has another negative repercussions, because we come into contact with the combination of early diagnosis and more severe cases, since such patients are referred with greater frequency to tertiary services, where the specialists are. These patients progress to premature fatal outcomes, reducing the overall cohort survival was the case with the sample analyzed in Minas Gerais,^{6,7} composed of 111 children, among whom around 1/3 of those diagnosed before 12 months of age died before completing that first year of life.⁶ To the extent that up to 2 years, and most especially during the first year after birth, our children attend basic health centers and they are cared by pediatricians and/or other professionals from the health teams, perhaps as many as ten visits, partnership with pediatricians would also make it possible to diagnose mild and moderate cases early too. It is opportune to point out here that we do not yet have estimates of under-reporting, we do not know the real number of undiagnosed CF patients. Undoubtedly, they number very much more than the 2,300 known cases.

Of course other strategies will always be welcome, but we understand that engaging pediatricians in cystic fibrosis care is one of them. In Brazil, as the authors point out, the number of specialized centers remains restricted and each one covers an extensive geographical area and has responsibility for large numbers of clients, which allows the assumption that there is a serious access problem for patients and their families with respect of these centers,¹ which might lead to late diagnosis.

Pediatricians have a strategic role to play in this context. Of course what we propose is a two-way referral and counter-referral system between pulmonologists, gastroenterologists pediatric nutrition specialists and general pediatricians. What then, in addition to adequate and continued training, both with physical attendance and distance learning, would be the pediatricians' activities in a possible reorganization of cystic fibrosis care? Among other actions, they should raise their level of diagnostic suspicion, think "fibro-cystically" when children have suggestive family backgrounds (abnormal sweat test, death from chronic respiratory diseases), meconium ileus, chronic and/or recurrent lung disease (in the awareness that within this group of diseases there are other. much more frequent, ones, asthma for example) and/or chronic diarrhea and/or malnutrition and/or failure to thrive and/or apparently inexplicable electrolyte disturbances and request sweat test in all cases presenting in this manner; promote health education, alerting parents and guardians to suspicious symptomatology and reinforcing basic notions of genetic counseling; participate in the clinical follow up of patients, assuming they feel comfortable doing so, in which case they

would work in conjunction with their peers at the closest specialized centers. We understand that, even though a distant reality in this country, some of these proposals are in agreement with those proposed by the Campinas team in this article and another, also published in the *Jornal de Pediatria*.⁸ To achieve this, it is indispensable that the number of laboratories capable of performing sweat test is increased in a responsible, measured and rigorously quality-controlled manner. Involving pediatricians implies providing them with the conditions that are necessary to perform this new function.

But would a care structure set out in this way truly be so far from the Brazilian reality? Perhaps not. In Belo Horizonte, during the last decade, there was just a single institution offering the sweat test and to this one, recently, four others have been added, both in the public (municipal, state and federal) and private sectors. In this manner, the supply of tests increased from around 1,000 to a minimum of 2,000 assays per year, in just five years and, as a direct consequence of this initiative, the Municipal Health Authority began distributing guidelines to pediatricians at primary health care centers stating clearly the findings that formally indicate sweat test. Furthermore, the Belo Horizonte inter-institutional agenda for CF, in 2002, included the implementation of a center to care for adults at the *Hospital das Clínicas* of Universidade Federal de Minas Gerais, one of the Brazilian institutions accredited for lung transplantation and, in 2003, the installation of a second pediatric reference center in the city also provided for is the institutionalization and continuation of this process of decentralizing care. Around five less complex facilities will be created for pediatric follow-up, equipped with resources for sweat testing, in the same number of key major cities around the State, duly interconnected and affiliated with the two institutions already implanted in the State capital. Clearly, it's possible to discern in all this the image of the implantation of a Statewide system, still faint it's true, but with potential multiplying effects and in no way ruling out the collaboration of general pediatricians since it will be brought closer to them.

Other recent movements on the Brazilian scene attest to an undeniable evolution in progress. Among these we would emphasize the creation, in 2003, of the Brazilian Cystic Fibrosis Study Group (*Grupo Brasileiro de Estudos em Fibrose Cística*) which coordinated the, now completed, guidelines for the condition, the reference document for the pediatric community and the Health Ministry. One inalienable role that this promising group has is to point out the imperative necessity for us to continue producing original data on the disease in our country, perhaps the reproduction across all Brazilian centers, and the totality of these centers as a set, of research such as the study here published by the Campinas group. In this motivating environment, the participation of the Brazilian Society of Pediatrics and its affiliates, aiming at links at federal, state and municipal levels, and even involving the various communication media (printed, spoken, televised) in educational campaigns about cystic fibrosis.

Last, but not least, we should continue producing a genuine body of knowledge about CF, as the authors of the article¹ so clearly pointed out, including analyses of adult patient samples, two of which were published recently.^{9,10}

Returning to the start. Dear pediatricians: allow us to invite you on board. We're in the same boat and we need more rowers in this boat. A shared voyage, not always over calm waters, but, certainly, a gratifying one. Just as the progresses described here illustrate well.

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