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REVIEW

Problems in the organization of care for patients with adult congenital heart disease

Problèmes dans l'organisation des soins de la cardiopathie congénitale

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Summary The prevalence of congenital heart disease among adults in Europe, or in any country in Europe, is not known. This is due to a lack of agreement on the incidence of congenital heart disease, with estimations varying from four per 1000 births to 50 per 1000 births, and it is not known how many patients with congenital heart disease have died. Based on several studies that estimated and calculated the number of adult patients with congenital heart disease, the number of patients should be much higher than the number of patients that are actually seen in specialized centres throughout Europe. This implies that either a large proportion of adult patients with congenital heart disease do not receive appropriate medical care, or that the calculations and estimations are grossly wrong. A combination of the two is also possible. A substantial expansion of the number and size of specialized centres for adult congenital heart disease is advocated, but since setting up (and running) a service for this disease is a costly affair, and because uncertainty remains about the actual number of patients needing specialized care, this has been difficult to realize in most European countries in the past few years.

MOTS CLÉS
Cardiopathie congénitale ;
Chirurgie cardiaque ;
Réseau de santé

Résumé La cardiopathie congénitale est la principale étiologie des défects congénitaux. En terme d'organisation des soins, l'impact des cardiopathies congénitales dans les premières années de la vie a surtout été mis en avant. Avant l'ère de la chirurgie cardiaque, les cardiopathies congénitales constituaient la principale cause de décès de l'enfant, et seulement une minorité de ceux nés avec une cardiopathie congénitale complexe, survivaient et atteignaient l'âge adulte. Après l'avènement de la chirurgie cardiaque, par Lillehei en

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1953, une amélioration progressive du taux de survie a été obtenue pendant les décennies suivantes. Cela a conduit à la situation actuelle, où le nombre de patients atteints de cardiopathie congénitale atteignant l'âge adulte a augmenté de façon progressive. Avec le niveau actuel de soins, il est escompté que plus de 90 % des patients nés avec une cardiopathie congénitale survivront et atteindront l'âge adulte. La majorité de ces patients qui survivent et atteignent l'âge adulte après une chirurgie cardiaque n'ont pas bénéficié d'un traitement « à vie » : la majorité en effet gardent des anomalies cardiaques résiduelles. La nécessité de la poursuite de soins spécialisés après les années d'enfance a été soulignée depuis le début des années 1980. L'existence de réseaux de santé pour les patients adultes atteints d'affection cardiaque, les services usuels de soins aux cardiaques, ne sont pas adaptés à cet objectif, du fait du manque d'expérience et donc de compétence dans ce domaine spécifique. Cette observation a conduit à établir des programmes pour des unités spécialisées dans l'Europe entière. Certains de ces programmes ont débuté dans les années 1980, la majorité dans les années 1990 et le nombre de centres spécialisés est en augmentation constante. De plus, le nombre de ces centres tente à augmenter rapidement.

Congenital heart disease is the most common of all congenital defects. In terms of organization of care, the emphasis has always been on the impact of congenital heart disease in the first years of life: in the era before cardiac surgery, congenital heart disease was the most important cause of infant death and only a few of those born with complex congenital heart disease survived until adulthood. Since the introduction of cardiac surgery by Lillehei in 1953, a gradual improvement in survival has been achieved over the subsequent decades. This has led to a situation where the number of patients with congenital heart disease who reach adult age has gradually increased, and with the current level of healthcare it is expected that over 90% of babies born in developed countries with congenital heart disease will now survive until adulthood.

Most patients who survive into adulthood after cardiac surgery in childhood, are not 'cured' for life; almost all have residual abnormalities [1]. The need for continuation of specialized care after their childhood years has been emphasized since the early 1980s [2]. The existing healthcare networks for adult patients with cardiac problems — the regular cardiology services — are not equipped for this task, because of lack of training and exposure and therefore skills in this specific field [3]. Awareness of this issue has led to the establishment of programmes for specialized adult congenital heart disease (ACHD) care throughout Europe. Some programmes started in the 1980s, many in the 1990s, and the number of centres is still increasing. In addition, existing centres tend to grow, many of them rapidly [4].

In a recent survey, Moons et al. [5] identified 70 centres in Europe that could be labelled as centres for ACHD. Altogether, these 70 centres had some 130,000 adult patients in their care. The authors stated that this was only a fraction of the entire population of patients with ACHD, that this population is heavily under serviced in terms of available care at an adequate level and that many more centres — or much larger units — would be needed.

Establishing and running a unit for specialized ACHD care is, however, a costly affair, as these chronically ill patients with considerable morbidity [6], need a relatively large amount of ''doctor time'' and claim a fairly large proportion of health-care resources [7]. If we want to convince healthcare planners and boards (or directors of hospitals) of the necessity of investing in such a costly service, we need to answer a few basic questions. How many patients in total are involved? How many of these patients have complex ACHD, how many have ACHD of moderate severity and how many have mild ACHD? Which defects really require specialized tertiary referral ACHD care and which can be dealt with in regional hospitals by cardiologists? How many patients need no special cardiac follow-up at all? These are simple and fair questions, but are difficult to answer.

We do not know the incidence of congenital heart disease (i.e., how many patients are born per year with congenital heart disease in a specific population or country), because reports on this topic vary enormously from four per 1000 live births to 50 per 1000 live births.

We also do not know the prevalence of congenital heart disease (i.e., how many patients are alive with congenital heart disease in a population or country), because we do not know the starting point (the incidence) or how many patients have died. We are not aware of any country in which there is a population-based registry that is solid and detailed enough to answer these questions.

What should be considered as complex congenital heart disease? There is no uniformity in the definition that has been used in the various published studies, task force reports and position papers. For example, tetralogy of Fallot and atrioventricular septal defects — both fairly large diagnosis groups — are defects that are classified as severe or complex by some and as moderately severe by others. Neither is there consensus about which patient group actually needs highly specialized, tertiary referral care. There is not much discussion about really complex congenital heart disease (it is accepted that patients should be seen in a specialized centre]) and there is also a shared belief that truly simple lesions do not need specialized care. But what is the best option for the group of patients with moderately complex congenital heart disease? Regional care provided by the regular cardiology services near the patient, tertiary referral specialized care, or both options as shared care? And who should do a catheter-based intervention or a surgical proce-