Charity Funds Research to Improve Care for Children with Congenital Heart Disease

National heart charity Heart Research UK has been "helping hearts" for 45 years, by funding research into the prevention, treatment and cure of heart disease, and providing community grants for projects that encourage people to lead heart-healthy lifestyles. Now the charity is funding two new medical research projects that are focussing on providing better care and support for children who have congenital heart disease.

A functionally single ventricle heart condition, where only the left or right side of the heart has developed properly, is one of the most serious cardiac deformities encountered at birth[1].The conditions are lethal without treatment, and numerous heart operations over two to three stages are required to create a balanced circulation. However, whilst advances in medical and surgical care have transformed the prognosis for these children, the number of infants dying between these surgical stages has remained a concern, particularly during infancy. This has prompted the development of home monitoring programmes (HMP) to encourage early recognition of problems developing in these infants whilst living at home, to reduce the risk of potentially life-threatening events.

Most evidence relating to HMP has been limited to children born with an underdeveloped left ventricle (hypoplastic left heart syndrome), and there is a lack of evidence regarding their role in infants with right-sided defects, despite their similar risk of sudden unexplained cardiac death[5]. Studies have focused specifically on HMPs for infants with left-sided heart defects, utilising equipment to measure their oxygen levels and digital scales to record their weight on a daily basis. A study undertaken in Wisconsin, USA[4,6,7] used an historical control group (all infants with a poorly formed left heart who had standard discharge information and no monitoring at home) and compared the results of a later group (where all infants with a poorly formed left heart were enrolled into the HMP). However, the duration of the study was different between the historical group (four years) and the HMP group (14 months)[4] which made comparisons difficult; for example, four of nine deaths in the historical group were related to respiratory illness, which may have been related to a greater number of 'winter' seasons in this group due to the longer duration of collected data.

Subsequent publications from the Wisconsin team[7] have suggested significant survival benefits from HMP, but all have been non-randomised studies and have relied on historical matched controls such that it is difficult to exclude the influence of changes and improvements in surgical techniques, case selection and institutional learning curves. The evidence in terms of the HMP alone improving the outcome for infants could therefore be questioned. Other centres in Europe have recently presented positive findings of HMPs; however the studies have small numbers of infants in their groups. As yet, the results of these studies are unpublished[3] and so complete appraisal is not possible. Additionally, infants with incorrectly formed right hearts are not included in the research that has been conducted and published to date. More recently a randomised controlled trial around home monitoring for infants with complex congenital heart disease has commenced at the Children's Hospital of Philadelphia. The team at Birmingham Children's Hospital and Coventry University will be networking regularly with their Principal Investigator, Professor Medoff-Cooper, to discuss ongoing findings of the two similar studies.

Birmingham Children's Hospital has the largest programme for the treatment of functionally single ventricle heart conditions in the UK. A Heart Research UK-funded research project at Birmingham Children's Hospital will include all infants with single ventricle hearts (right and left), thereby increasing the number of infants who might benefit from this programme. It is the first prospective randomised study to assess the efficacy of HMPs in the UK. Additionally, this study will involve the education of parents to identify signs of early clinical deterioration in their infant and to use this 'early assessment tool' as part of the HMP. The study will also explore the impact that an HMP has on parental anxiety, depression and confidence, and the impact of these as well as parental demographics on the transition from hospital to home.

The research will test the feasibility of using a Congenital Heart Assessment Tool (CHAT) as part of a home monitoring programme (HMP) for infants with single ventricle heart conditions. The HMP includes daily measurements by parents of their infant's oxygen levels and weight, whilst discharged home between the first and second stages of treatment. All the equipment for the families is provided by the Heart Research UK grant. Additionally, the CHAT uses a traffic light system to give an early indication of deterioration of the infants' condition, helping to inform parents and medical teams promptly about problems that may be occurring so that the infant can be treated as soon as possible. The HMP/CHAT has been developed by the researchers, clinicians and an expert advisory group. The study will now recruit a group of parents and infants to test and feed back on the suitability of the discharge strategy. The recruited parents and infants will be separated into three groups for the research project: those using the HMP/CHAT, those using only the CHAT, and a third group receiving only the current standard NHS discharge procedure.

The overall aim of the project is to determine whether the CHAT/HMP is suitable and of benefit to parents and infants, and to provide data to inform the development of a larger-scale trial. Data provided will help define whether or not lives can be saved by

the HMP, and establish whether the CHAT individually or the CHAT with HMP best aids early recognition of deterioration of the infant's condition. It also aims to examine how confident parents are and whether the tools can help alleviate parental anxiety and depression.

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Access to Clear Advice about Exercise for Children and Adolescents with Congenital Heart Disease

Congenital heart disease (CHD) includes numerous structural and functional abnormalities of the heart, each of which may occur in isolation or in conjunction with other defects. Therefore, patients described as having CHD represent a heterogeneous group, varying greatly in their treatment, prognosis, exercise capacity and quality of life. The occurrence of CHD is estimated at 1 in 145 births[8]. Considerable advances in medical intervention have resulted in significant improvements in prognosis, with 85% survival to adulthood[9]. This has led to a paradigm shift within healthcare, from focusing almost exclusively on patient survival to a broader consideration of quality of life[10].

The majority of people with CHD have a reduced capacity for exercise. This can be quantitatively assessed by cardiopulmonary exercise testing, which measures parameters such as peak oxygen consumption (VO2peak) and heart rate reserve. For individuals with severe defects, exercise intolerance may be caused by cyanosis, pulmonary hypertension, or a limited ability to increase cardiac output. In the case of mild defects, it may simply reflect deconditioning due to an inactive lifestyle. Poor exercise tolerance is linked to poor prognosis[11].

However, a number of studies have demonstrated significant improvements in the exercise capacity of young people with CHD following exercise training[12]. These benefits can be sustained for a prolonged period following rehabilitation[13].

Many people with CHD can safely participate in sports and physical activities with minimal restrictions. Indeed, recent recommendations state that, in general, children with CHD should be encouraged to comply with public health guidelines of ≥60min of moderate to vigorous activity each day[14]. However, for some patients, certain activities may be harmful or indeed life-threatening. It is therefore, essential that clear advice is given. Several consensus documents have recently been published, which provide exercise recommendations for specific heart defects[14,15,16]. Whilst these give a useful framework for clinicians, advice about exercise should be individualised; taking into account the patients' underlying pathology, their age and sporting interests. Ideally cardiopulmonary exercise testing would also be used to determine the safe limits of exercise intensity.

The benefits of exercise for children and adolescents have been well documented, and include improvements in: musculoskeletal development, social interaction, psychological wellbeing and weight management[17]. These advantages apply equally to young people with CHD and their healthy peers. In specific defects, there may be additional reasons for prescribing exercise. For example, some conditions increase the risk of premature atherosclerotic cardiovascular disease, i.e., congenital coronary artery abnormalities, coarctation of the aorta, and patients who have received an arterial switch operation[18]. For these individuals it is particularly important to avoid becoming overweight or obese[19], since this is an important risk

factor for atherosclerosis. However, a recent study found the prevalence of overweight or obesity to be 27% within the paediatric CHD population[20].

Despite the clear benefits of exercise, several studies report a tendency towards a sedentary lifestyle in people with CHD, irrespective of their capacity[21]. It has been suggested that this is due to anxiety and overprotection on the part of the parents. It also seems likely that healthcare professionals and schools will err on the side of caution, preferring to advise against exercise rather than risking adverse consequences. However, national differences in healthcare provision, access to sporting facilities and social attitudes may invalidate conclusions based on data collected in another country. Therefore, the true extent of the problem within the UK remains unclear.

For most children and adolescents the ability to participate in sporting activities, particularly within the school environment, plays an important role in social integration and self-perception. Clearly it is important that young people gain a clear understanding of which types of exercise are appropriate for them. Unfortunately, there is often some confusion in this area. As a result, many young people are restricted in their participation by uncertainty and anxiety concerning their heart condition. More worryingly, a significant proportion of young people believe themselves to have no exercise restrictions, when in reality their condition makes it dangerous for them to participate in certain activities[22]. In the absence of clear information, patients tend to assume that all exercise is safe for them. Their parents, however, often assume that most exercise is best avoided. There is an urgent need to improve communication in this area, in order to ensure that all young people gain the maximum benefit from leading an active life, whilst keeping within safe parameters.

Heart Research UK is funding a project to investigate the provision of exercise advice to young people with CHD. Based at the Children's Health and Exercise Research Centre, University of Exeter and Bristol Royal Hospital for Children, the project will assess the current availability of appropriate advice and rehabilitation services in the UK, and evaluate practice across the regions.

It also aims to gain a clearer understanding of patients' knowledge regarding the type and intensity of sport which they can safely enjoy. This information will be used to inform the production of an information resource pack for children and adolescents with CHD, their parents and other care professionals. This resource will help to bridge the gap in knowledge, highlighting the benefits of exercise and emphasising what patients can do.

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