

Long-term Outcomes

Adaptation of the child and family to life with a chronic illness

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CHRONIC ILLNESS IN A CHILD PRODUCES STRESS for both the child with the illness and the family of which he or she is a part.¹ Today, it is estimated that greater than one-tenth of children are living with some form of chronic illness or condition.^{2–3} Faced with this stress, children and families are required to adapt to potential physical, emotional, social, and financial challenges. Professionals providing health care have an opportunity to influence how children and families interpret and adapt to these challenges. Guidance can be drawn from the multiple theoretical perspectives that have explored the process of adaptation to chronic illness.

Congenital cardiac malformations are amongst the conditions that require adaptation by both the child and family. The American Heart Association estimates that 1 out of every 125 infants are born with some form of congenital cardiac disease, making it the most commonly reported form of structural birth defect.⁴ Despite dramatic improvements in medical and surgical management, these conditions remain the leading cause of death in infants related to congenital malformations.⁵ The affected child is not the only one that experiences the impact of the diagnosis of a congenital cardiac malformation. Children and families may feel that they are riding a roller coaster that includes uncertainty about long-term prognosis, chronic need for medication, repeat interventions, persistent symptoms, and the prospect of physical and/or developmental delays. Families facing this crisis are in need of support, education, and understanding in order to help them initially adjust, and ultimately reach a level of adaptation to this challenge.^{1,6–7} Little

information is available on how children and families adapt to living with these issues, or the strategies used by the families to support their integrity. Several questions remain unanswered. Why do some children and families appear to adapt easily while others do not? What are the risk factors for poor adaptation? Are there interventions that could improve adaptation for children and families? Do children, mothers, and fathers experience the same or different trajectories of adaptation?

The purpose of my review is three-fold. First, I will review selected theoretical approaches to the study of child and family adaptation to chronic illness. Second, I will summarize existing findings on adaptation of the child and family in response to the diagnosis of a congenital cardiac malformation. Finally, I will suggest directions for further research and intervention in this area. The review, hopefully, will help providers of health care to define the implications of chronic illness for the child and family, specifically congenital cardiac disease, to identify risk factors for poor outcomes, and to provide a guide for the development of interventions to improve the well-being, adaptation, and overall quality of life of both the afflicted children and their families.

Adaptation of the family

The response of families to the diagnosis of a chronic illness in their children is a complex and multifaceted process.^{1,8–9} McCubbin et al.⁸ suggest that this process occurs in stages, with multiple intervening variables. Initially, families experience a period of adjustment, involving early responses and changes to cope with the stress of the diagnosis of chronic illness, a parent, for example, taking a leave of absence from employment. Over time, most families progress to a state of

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long-term adaptation to the demands of the chronic illness suffered by their child, demonstrating the inherent resiliency of the human spirit.⁸ Austin¹⁰ (p. 104) defines adaptation as “any response to the chronic illness by a family member”. The response may be positive or negative. Adaptation is the process of reducing discrepancies between expectations and reality in order to reduce psychological distress.⁸ Examples of long-term adaptation may include tangible changes, such as moving to a home equipped to deal with the handicap, or incorporating the administration of medications into daily routines. Successful adaptation is also manifested by psychological functioning, including self-acceptance, positive relations with others, autonomy, a feeling of purpose in life, environmental mastery and the potential for continued personal growth.¹¹

Systematic investigation of the process of adaptation of the family to the stress of chronic illness in a child provides evidence on which to base structured interventions, to support healthy adaptation, and to identify families at risk for maladaptation. Several researchers have examined the phenomenon of adaptation of the family to the presence of chronic illness in a child.^{8,12–15} Over time, most families do demonstrate successful adjustment and adaptation to this type of stress. When this does not occur, both the affected child and the family unit are at risk for consequences related to maladjustment.¹ These consequences are not limited to, but may include, psychological illness, negative effects on quality of life for the child and/or family, marital distress, and negative impact on siblings. Family adaptation is not a static entity. Members of the family may move in and out of periods of successful adaptation depending on the demands imposed by the illness.¹⁶ It is not uncommon for different members to manifest varying degrees of adaptation.¹²

Several conceptual models and theories have been proposed to describe how a family seeks the restoration of balance and harmony in response to stress, and what factors influence the eventual outcome.^{8,13,17–18} Many of these models have been adapted from the theory of stress and coping as described by Lazarus and Folkman.¹⁹ Psychologists, specialists in child development, family theorists, and nurses are some of the specialists that have explored this concept. Each of these disciplines offers unique perspectives and insights into this phenomenon.

The theory proposed by Duvall¹⁷ emphasizes the evolution of a family over time. Change in families is initiated in response to the developmental tasks being faced, and the stress associated with progress through each critical period of development, for example the various stages of childrearing. Given today’s diverse definition of “family”, few families will strictly follow

the sequential progression through the stages that Duvall originally described. This theory, however, identifies some useful variables for consideration when assessing a family faced with the challenge of chronic illness in a child. These include the age or the level of maturity of the family members involved, the duration of their relationships with each other, the marital state of the parents, and the birth order of the child affected by chronic illness. The theory of family development asserts that the stress of the diagnosis of chronic illness in a child must be considered within the context of the normal challenges that a family may be facing.

Systems theory, and social ecology, emphasize the social context of the child and family as the critical factor influencing the processes of coping and long-term adaptation.^{13,15} The nature and quality of interactions with peers, the school system, parental social support, and systems of community support are all believed to contribute to outcomes. This model, based on systems theory and social ecology, expands the framework of variables proposed to influence adaptation by the family to the diagnosis of chronic illness in their child beyond that proposed by the theory of family development alone. These theories address the chronic phase of the illness, however, they place little emphasis on the perception of the initial stress. The internal resources of the family, and their appraisal of the stress, will also have an important impact on the adaptation of the family.

In the Resiliency Model of adjustment and adaptation of the family,⁸ several theories of familial response to stress have been refined to produce a complex model outlining the multiple factors influencing the outcome for the family. Resiliency, defined as, “the positive behavioural patterns and functional competence individuals and the family demonstrate under stressful circumstances, which determine the family’s ability to recover by maintaining its integrity as a unit while insuring, and where necessary restoring, the well-being of family members and the family unit as a whole”,⁸ represents the driving force that moves a family through the process of adaptation. The model proposes that highly resilient families are more likely to progress quickly, and have more favourable outcomes, than less resilient families. Several variables will impact on this process.^{20–21} Variables for the child include age, gender, birth order, developmental stage, and the ability to comprehend the condition and its treatment. Variables related to the condition include the age of onset, level of severity, prognosis, predictability of symptoms, nature of the limitations imposed, and effects of the treatment. Variables for the family include the developmental stage, their prior experience with stress, the state and trait anxiety of individual family members, inherent family hardiness,

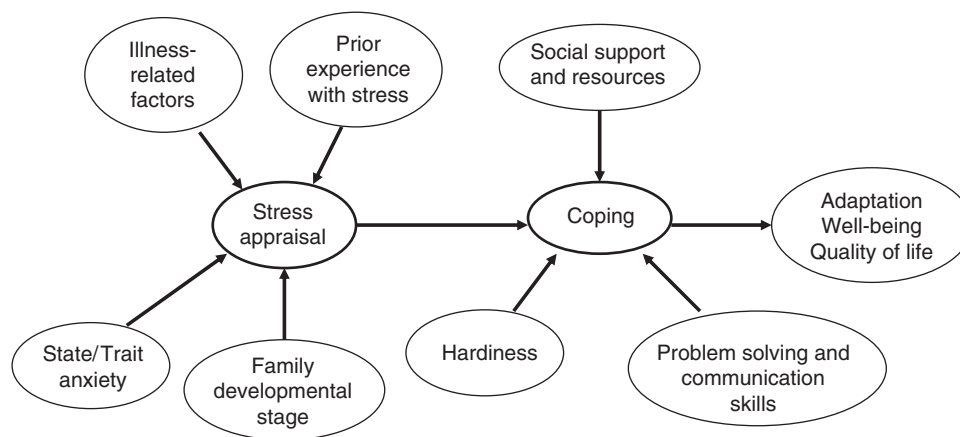


Figure 1.

Adaptation by the family to chronic illness in their child. (Adapted from McCubbin, Thompson, and McCubbin, 1996;⁸ Leske, 2000;²⁰ and Leske, 2003.)²¹

established problem-solving and styles of coping, socioeconomic state, and available systems for social support, as well as religion and culture. A model can be proposed to show the relationships between these variables (Fig. 1). The initial perception of the stressor by the child and family will have a considerable impact on their ability to adapt. For example, families that have successfully coped with challenges in the past will be able to draw from prior experiences, and may find strength in their demonstrated ability to “get through things”. In contrast, a family that has very high expectations for their experience as parents, and little history of coping with stress, may perceive the diagnosis of chronic illness in their child as an insurmountable challenge.

Existing theories, therefore, highlight the multiple variables influencing the responses of the child and family to the diagnosis of chronic illness. The severity of the illness is one factor, but does not independently predict how a child and family will respond. When assessing the ability of a family to adapt, all of the aforementioned factors must be taken into consideration. Interventions to support resiliency, and promote positive styles of coping, may aid successful adaptation.

The impact of the congenital cardiac malformation

The psychosocial outcomes of children and families living with congenital cardiac disease are receiving increased attention. Survival has become the expected outcome for most affected children. As a result, interest has shifted to exploring longer term results for both the child and family. Studies available to date have been primarily descriptive in nature, typically including samples characterizing the experience of a single centre, and often representing subjects with very heterogeneous diagnostic backgrounds. These facts have

limited broad generalizations about psychosocial outcomes, but despite these limitations, some important findings exist to guide future research.

Parents of children with congenital cardiac disease have been found to experience more anger and sadness than parents of healthy children.⁶ They have also reported more social problems, less leisure activity,²² greater feelings of distress and hopelessness⁷ and higher levels of overall stress^{23–25} than parents of healthy children, or those of children with other chronic conditions such as cystic fibrosis. Mothers and fathers have differed in what they report as the sources of stress in caring for infants with congenital cardiac disease.²⁶ Utens et al.^{27–28} have studied extensively the psychological responses of children and parents to congenital cardiac malformations. Parents, especially mothers, of children awaiting cardiac intervention reported elevated psychological distress, and less adequate styles of coping, compared to reference groups.²⁷ This response was independent of the age of the child, and parental reports of stress decreased after intervention.²⁸ In a sample of 80 parents, Uzark and Jones²⁵ reported that 17.5% reported parental stress greater than the 90th percentile on the Parenting Stress Index. Level of stress was unrelated to the severity of the malformation. In a study exploring neurodevelopmental and psychosocial outcomes in preschool-age survivors of transposition and hypoplastic left heart syndrome,²⁹ a strong positive correlation was identified between behavioural problems and parental stress (Fig. 2). Behavioural problems, including attention deficit disorder, have surfaced as one of the more common co-morbidities in children with congenital cardiac disease.^{30–31} Whether these problems have an organic nature, result from environmental influences, or are a result of psychological response of the child to the stress of chronic illness, remains unknown.

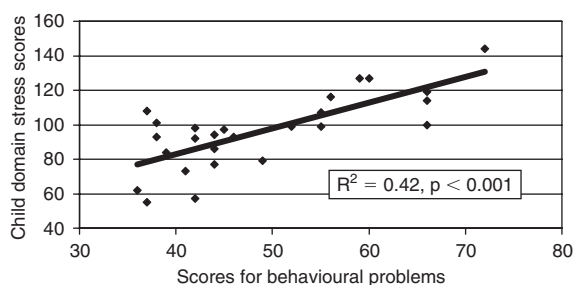


Figure 2.

Behavioural problems in the child, and parental stress, are positively correlated ($r = 0.65$, $p < 0.001$). Total scores for behavioural problems explained 42% of the variation in child domain stress scores. The sample was made up of 13 children with hypoplastic left heart syndrome, with a mean age of 3.9 years, and 13 children with transposition, their mean age being 4.6 years. Child domain stress scores from Parenting Stress Index. Total scores for behavioural problems from the Child Behavior Checklist.

One study has specifically addressed the psychosocial implications of hypoplastic left heart syndrome. Thus, Williams et al.³² identified significant uses of resources in this population, which can be translated to financial impact for families, and negative effects on quality of life for both the children and their parents. Positive outcomes for the family may also be possible. In a survey of 209 parents, Wray and Maynard³³ found that over nine-tenths reported stable or improved family cohesion, and over one-third reported improved relationships between partners.

Psychosocial adaptation of children living with congenital cardiac disease has also been explored, and similarly, both positive and negative implications have been identified. Children have reported poorer body image and self-concept, as well as greater anxiety and incidence of behavioural problems, than healthy children.^{34–35} Symptoms of post-traumatic stress disorder have been identified in children after surgical repair of cardiac defects.³⁶ There have been mixed results on the impact on quality of life for children living with congenital cardiac disease, with some research reporting negative impact on quality of life, and others reporting quality of life equal to healthy peers.^{37–39} Positive findings have included lower anxiety, and greater superego strength, in adolescents with congenital cardiac disease compared to healthy peers.⁴⁰ Utens and colleagues found behavioural and emotional functioning in young adults with congenital cardiac disease to be similar to a healthy reference group.⁴¹

The only consistent finding regarding adaptation of the child and family to congenital cardiac disease has been that severity of the malformation is not a reliable predictor of long-term psychosocial outcomes.^{25,32,41,42} In a revealing study, DeMaso et al.⁴³ explored the impact of maternal perceptions regarding the diagnosis of the disease on adjustment of the affected

child. One-third of the variability in adjustment by the child was explained by maternal perceptions of the disease, compared to only 3 percent explained by medical severity. The “life and death” nature of diseases associated with the heart may account for responses from parents or children, which at times seem out of proportion to the diagnosis.^{25,32} Although sometimes seeming illogical to health care professionals, the perceptions and responses of any given child or family are real, and deserve attention. Physicians and nurses providing practical guidance, and consistent communication, about the implications of congenital cardiac disease may assist families in developing accurate perceptions of the impact of this diagnosis.

Congenital cardiac disease represents a chronic condition for the majority of survivors, despite dramatic improvements in medical and surgical care. Children living with such disease, and their families, must adapt to the challenges of this diagnosis over a lifetime. Follow-up visits to physicians, repeat intervention, changes in therapy, or transition to different caregivers, may be particularly stressful. Children may report different effects of the disease as they progress through normal developmental stages. For example, a child who underwent a single surgical repair as an infant may not be fully cognizant of his or her condition until they become aware that other children do not have a sternotomy scar. Similarly, the perceptions of personal vulnerability, or the stress of limitations of activity, will be very different for a five year old as compared to a fifteen year old. Families will also experience varying degrees of impact according to their developmental stage, current burden of stress, and the results of their attempts to cope.

Implications for practice and further research

Caring for children and families with chronic conditions, such as congenital cardiac malformations, requires attention not only to their physical health, but also to their psychosocial health. To do this, health care professionals must be perceptive and empathetic to the stress of chronic illness. Just as we assess risks for the child with cardiac disease, we should also assess the family for risks of problems with adaptation to this challenge (see Appendix). Asking, “How are you doing?” is not enough unless we are prepared to listen actively and intervene based on the response. We must inform families of expectations for their child, taking care to be honest about what we know, and especially about what we do not know. Mechanisms for long-term psychosocial support for children and families need to be in place within our cardiac centres. These services should include social workers, psychologists, and patient advocates.

Further research should include longitudinal studies on adaptation by the child and the family to the diagnosis of congenital cardiac disease across the lifespan. Knowledge gained from this type of inquiry will allow caregivers to plan better for developmentally appropriate support. Screening tools for assessment of risk need to be developed to identify children and families at risk for poor adaptation. Screening should include all of the variables related to the child, the family, and the condition that have been demonstrated to impact on adaptation. This screening should occur at the time of initial diagnosis, and continue throughout the course of treatment for a child and family. Given prior results, the assumption that severity of illness is the primary predictor of long-term adaptation must be abandoned. Instead, we must explore the subjective perceptions of children and families, and assist them in developing a realistic assessment of the impact of congenital cardiac disease on their lives. Ultimately, interventions to promote resiliency, and effective strategies for coping, need to be designed and tested. There is no “one size fits all” intervention to support adaptation for children

and families. It is likely that tailored interventions, designed to meet the individual needs of specific children and families, will be most successful. These may include training in the skills of coping, promotion of self-efficacy, techniques to reduce stress, and other strategies.

Conclusion

Children and families exhibit a complex response to the stress of chronic illnesses like congenital cardiac disease. Adjustment and adaptation to this stress are processes that are influenced by multiple variables. A thorough understanding of these variables is required to assess risk for poor adaptation. Children and families are naturally resilient, but successful adaptation requires ongoing support and understanding. Both positive and negative outcomes are possible. Caregivers of children and families living with chronic illness have an opportunity, and an obligation, to promote successful adaptation through further research and intervention.

Appendix

Assessment of risk for the family

Assessing families for risk of problems with adaptation to the stress of chronic illness in a child requires consideration of multiple variables concerning the child, the family, and the illness. As an example, consider two hypothetical families both faced with the diagnosis of hypoplastic left heart syndrome in a newborn.

Family # 1:

- Married one year, white, suburban couple
- 1st child
- 2 careers, both college graduates
- Own a home
- Good insurance
- Prenatal diagnosis
- Extensive family and community social support system

Family # 2:

- Single, minority mother
- 3rd child
- Unemployed, high school graduate
- Rents an apartment
- On public assistance
- Postnatal diagnosis
- Social support available but not always reliable
- Maternal grandmother of infant provides childcare

Which family is at greater risk for problems with adaptation?

Although the single mother in the second family faces several important social risk factors that require attention, in some ways her situation may have prepared her better for adaptation to life with this new infant affected by complex congenital cardiac disease. She has extensive prior experience with stress, and has had to learn how to cope with adversity. She has experience as a mother that she will draw upon as she learns to parent this child, and meet his or her special needs. She has learned how to access the systems providing social support to meet her basic needs, and will likely seek out guidance from a social worker on how to extend these services to assist with the needs of this child for instance, transportation to and from frequent medical visits. The consistent and strong presence of the maternal grandmother will be a major source of guidance and support.

At first glance, the first family may appear to be able to handle anything, however, they may be the

family at greatest risk for problems with adaptation. Although married, their relationship is young, and they have not had the chance to develop their roles as parents. Prenatal diagnosis allowed them to gather information about what they would face, albeit that knowledge about risks and outcomes may affect their ability to bond with this child. They have had little prior experience with stress, and may not have developed adequate skills for coping with challenges. Both parents have career interests, and although they have good insurance, they also have a home mortgage to pay. It is likely that one parent will have to sacrifice his, or her, career during the early months of the life of their child, life which may impose significant financial burden, as well as being resented by the parent who had to alter his or her plans. The system for social support to which they are connected may rally initially, but may find it difficult to address the needs of a child with a complex chronic illness. The parents may find themselves feeling isolated, yet unwilling to access structured social support because of the stigma they associate with it.

Providing routine psychosocial assessment and follow-up by a trained care provider may help to reduce problems with adaptation. Families should be counselled that there is significant stress associated with caring for a child with special needs, and that accepting help is not a sign of failure. Providers of care must listen to the concerns of the family, and take time to be perceptive and empathetic. The human spirit is incredibly resilient, and most families will adapt successfully, albeit that this requires guidance and support.

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