**ABSTRACT**

In an effort to provide counseling for families, we reviewed long-term outcomes in patients with Noonan syndrome (NS) and cardiomyopathy (CM). In a cohort of 47 patients with CM diagnosed in infancy, we determined their survival, age at diagnosis, gender, and race/ethnicity. The Pediatric Cardiology Registry (PCR) was sponsored by the American Heart Association, and has contributed to the understanding of cardiac disease in children. The PCR cohort included 31 children with Noonan syndrome (NS), a genetic disorder characterized by a distinctive facial appearance and a variety of cardiovascular anomalies. Since the first description of Noonan syndrome (NS) in 1963, more than 400 patients have been described. The clinical features of Noonan syndrome include distinctive facial features, short stature, cardiac anomalies, and developmental delay. Noonan syndrome is a genetic disorder, and genetic testing has been performed in many cases. The PCR cohort included children with Noonan syndrome who were diagnosed with CM between 1980 and 2001, and on initial presentation. The cohort was followed until death, transfer, or the age of 18, whichever occurred first. The cohort was followed prospectively (1980-1985), with annual follow-up (1985-1996). Despite advances in genetic testing, Noonan syndrome remains elusive. The PCR cohort includes patients with Noonan syndrome and cardiomyopathy, and their outcomes and survival have been documented.

**METHODS**

The PCR enrolled 62 patients who were diagnosed with hypertrophic cardiomyopathy between 1991 and 2000 as part of a cohort study of patients with CM. The PCR cohort included 31 children with Noonan syndrome (NS), a genetic disorder characterized by a distinctive facial appearance and a variety of cardiovascular anomalies. Since the first description of Noonan syndrome (NS) in 1963, more than 400 patients have been described. The clinical features of Noonan syndrome include distinctive facial features, short stature, cardiac anomalies, and developmental delay. Noonan syndrome is a genetic disorder, and genetic testing has been performed in many cases. The PCR cohort included children with Noonan syndrome who were diagnosed with CM between 1980 and 2001, and on initial presentation. The cohort was followed until death, transfer, or the age of 18, whichever occurred first. The cohort was followed prospectively (1980-1985), with annual follow-up (1985-1996). Despite advances in genetic testing, Noonan syndrome remains elusive. The PCR cohort includes patients with Noonan syndrome and cardiomyopathy, and their outcomes and survival have been documented.

**RESULTS**

The PCR cohort included 31 children with Noonan syndrome (NS) and 31 children with isolated cardiomyopathy (CM). The cohort was followed until death, transfer, or the age of 18, whichever occurred first. The cohort was followed prospectively (1980-1985), with annual follow-up (1985-1996). Despite advances in genetic testing, Noonan syndrome remains elusive. The PCR cohort includes patients with Noonan syndrome and cardiomyopathy, and their outcomes and survival have been documented.

**DISCUSSION**

Since the first description of Noonan syndrome (NS) and cardiomyopathy by Drs. Noonan and Emhke, the pediatric cardiomyopathy literature has continued to grow. The discovery of hypertrophic cardiomyopathy (HCM) in patients with Noonan syndrome has raised the question of long-term management strategies. Although outcomes and survival data are limited, the problem is defining who is at high risk. Rising incidence and earlier age of identification of Noonan syndrome have a Pediatric Cardiology Registry (PCR) that can provide a certain profile predictive of increased risk. In an effort to provide counseling for families, we reviewed the outcomes and survival of patients with Noonan syndrome and cardiomyopathy. The large cohort size and population-based design of the PCR cohort allows for a more comprehensive understanding of the outcomes and survival of patients with Noonan syndrome and cardiomyopathy. The Discovery of hypertrophic cardiomyopathy (HCM) in patients with Noonan syndrome raises the question of long-term management strategies. Although outcomes and survival data are limited, the problem is defining who is at high risk. Rising incidence and earlier age of identification of Noonan syndrome have a Pediatric Cardiology Registry (PCR) that can provide a certain profile predictive of increased risk. In an effort to provide counseling for families, we reviewed the outcomes and survival of patients with Noonan syndrome and cardiomyopathy. The large cohort size and population-based design of the PCR cohort allows for a more comprehensive understanding of the outcomes and survival of patients with Noonan syndrome and cardiomyopathy. The Discovery of hypertrophic cardiomyopathy (HCM) in patients with Noonan syndrome raises the question of long-term management strategies. Although outcomes and survival data are limited, the problem is defining who is at high risk. Rising incidence and earlier age of identification of Noonan syndrome have a Pediatric Cardiology Registry (PCR) that can provide a certain profile predictive of increased risk. In an effort to provide counseling for families, we reviewed the outcomes and survival of patients with Noonan syndrome and cardiomyopathy. The large cohort size and population-based design of the PCR cohort allows for a more comprehensive understanding of the outcomes and survival of patients with Noonan syndrome and cardiomyopathy.