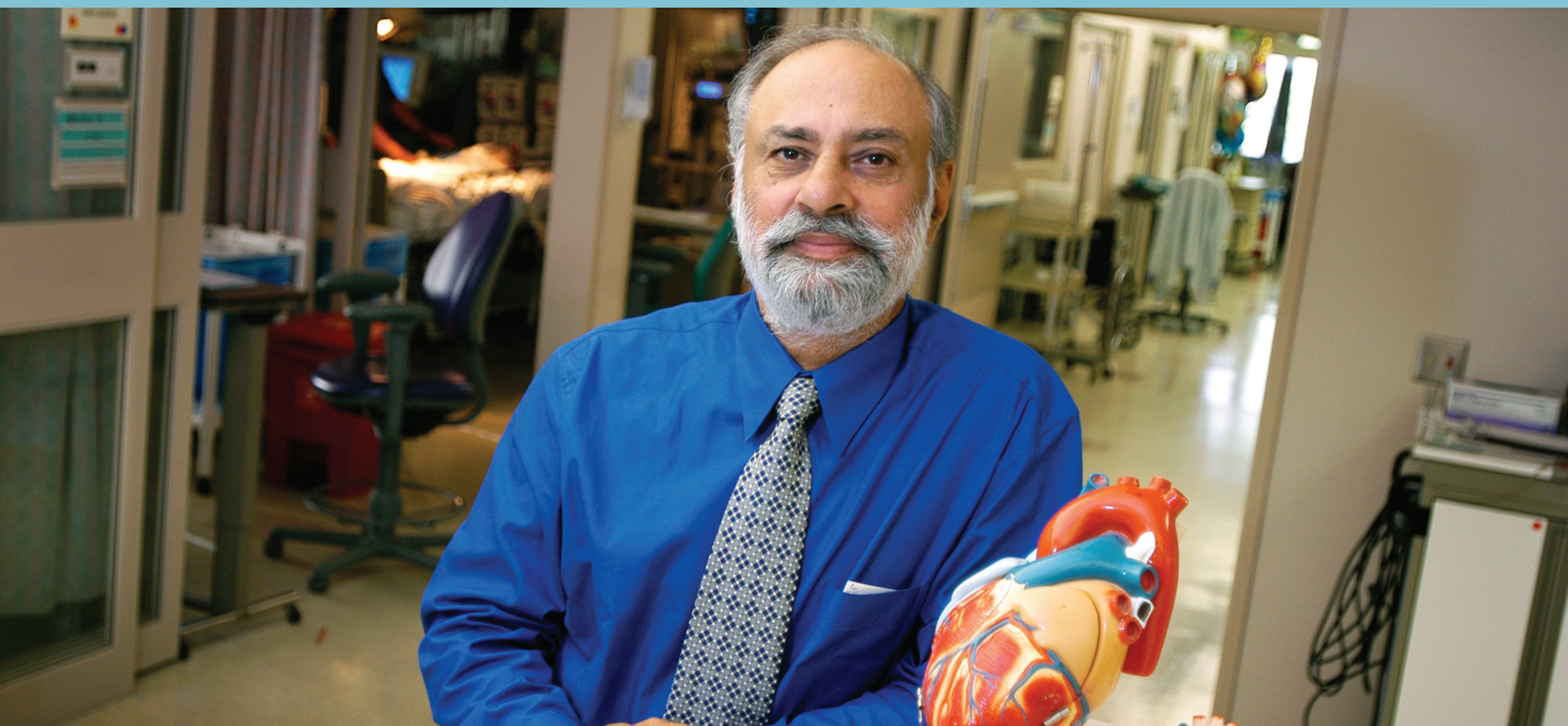


Until the 1980s, the congenital heart defect called hypoplastic left heart syndrome was a fatal diagnosis. Children's has been a pioneering hospital in a series of surgeries to treat this defect. Amarjit Singh, MD, studied 20 years of outcomes among Children's patients who had these surgeries.



Pediatric cardiologist Amarjit Singh, MD, in the pediatric intensive care unit at Children's – Minneapolis.

Children's of Minnesota cares for nearly two-thirds of pediatric patients hospitalized for cardiac surgery among hospitals in the Twin Cities metropolitan area.

Twenty-Year Experience With Hypoplastic Left Heart Syndrome In a Free-Standing, Community-Based Children's Hospital

Principal Investigator:

Amarjit Singh, MD, Pediatric Cardiologist

Co-Investigator:

David Overman, MD, Pediatric Cardiac Surgeon

Introduction

This study reviewed 20 years of treatment for patients with hypoplastic left heart syndrome (HLHS) at Children's Hospitals and Clinics of Minnesota and the Children's Heart Clinic. HLHS is one of the most serious congenital heart defects, affecting up to four of every 10,000 live births. It is among the top three heart abnormalities necessitating surgery in newborns.

Methods

Patients studied were admitted to Children's of Minnesota with the diagnosis of HLHS since the treatment program's inception in 1985. Analysis by era in five-year increments (1985–1990; 1991–1995; 1996–2000; 2001–2005) was performed for demographics, election for surgical therapy, early and late outcomes, prenatal diagnosis, and anatomic and procedural factors. In all, 60 data elements were considered.

Demographic, anatomic, and procedural factors affecting survival were sought, using univariate and multivariate analysis.

Outreach efforts by Singh and other physicians in the Children's Heart Clinic resulted in referrals of newborns with suspected heart disease by physicians from Greater Minnesota, northern Iowa, western Wisconsin, and North and South Dakota.

Results

One hundred eighty-four patients were identified. Demographic variables were uniform over the four five-year eras. Of the group, 144 elected to undergo surgical therapy. For the entire series of three surgeries to treat HLHS, early survival by stage was: 68 percent for Stage I; 88 percent for Stage II; and 94 percent for Stage III. These results did not change significantly when analyzed by era or surgeon.

Further analysis found cumulative survival for Stages I and II was significantly improved for patients in the final era, 2001–2005. Mortality for the entire series was 12 percent between Stages I and II and 14 percent between Stages II and III. Interstage mortality decreased significantly in 2001–2005.

Discussion

Successful surgical treatment of HLHS was achieved during the study period at Children's. These results mirror published results of care for HLHS at other leading institutions. Cumulative survival has improved significantly when the most recent era is compared with prior eras.

Singh's First Patient to Undergo Norwood Surgery Prompts 20-Year Review

When Amarjit Singh, MD, started his practice at Children's Heart Clinic in 1975, babies born with hypoplastic left heart syndrome (HLHS) died soon after birth. Ten years later, an infant with this congenital heart defect became a turning point in Singh's career.

Singh and his colleague, the late cardiac surgeon Hovold Helseth, MD, had followed the progress of William Norwood, MD, a pioneering surgeon at Children's Hospital of Philadelphia. Norwood pioneered a surgery in 1983 to rebuild the heart of an infant with HLHS soon after birth.

In April 1985, a Wisconsin family whose infant son was born with HLHS called Singh for help. The baby, Doug Smieja, was transferred to Children's. Singh asked parents Kathy and Gary Smieja whether Helseth could try the new surgery. They agreed, understanding that although the surgery was risky, Doug otherwise had no chance to live.

This was the first time the Norwood surgery had been performed at Children's of Minnesota and in the Upper Midwest. Children's joined a small group of pediatric hospitals in the U.S. to pioneer the three-stage surgical treatment for HLHS.

The surgery saved Doug's life. Under Singh's care, he went on to have the Glenn and Fontan surgeries when he was older.

Smieja, who still receives care from Singh, celebrated his 20th birthday in 2005. That milestone prompted Singh to research outcomes for 184 HLHS patients cared for at Children's from 1985 to 2005. Singh says he was pleased to find that the results at Children's matched those of the top centers for pediatric cardiology in North America.

Singh presented his research in 2006 to the Midwest Pediatric Cardiology Society, including the tremendous progress in care that Doug Smieja represents.

"It's very gratifying to see these kids grow up and have a good quality of life," Singh says. "Good outcomes are a credit to our team approach of caring for these patients."