Treatment for Facial and Internal Hemangiomas of Patient with PHACE Syndrome

Complex condition complicated by severe coarctation, absence of carotid and vertebral arteries

Clinical History and Presentation

In 2007, the parents of a newborn female diagnosed with PHACE syndrome brought their child to the Hemangioma and Vascular Malformation Center at Cincinnati Children’s Hospital Medical Center after being told by their local hospital that their daughter was not a candidate for surgery and they should seek hospice care. Upon receiving this news, the parents sought a second opinion. They researched available options online and spoke with representatives of the National Organization for Vascular Anomalies (NOVA), who recommended Cincinnati Children's for the second opinion.

The child met the criteria for definite PHACE syndrome: severe coarctation of the aorta, absence of her left carotid and vertebral arteries, and a diffuse or segmental hemangioma on her face.

Our Approach

Denise Adams, MD, medical director of the center, was the first physician to examine the child at Cincinnati Children's and plot a course for treatment, which involved taking into consideration the numerous complex medical conditions with which the patient presented. Dr. Adams became the coordinator of the child’s care throughout the long process that was to follow.

At the child’s first visit, Dr. Adams ordered a blood transfusion after blood work showed the child’s hemoglobin to be 6.7, suggesting breakdown of the red blood cells (hemolysis). An ultrasound was done, revealing a mass around her spleen and abdomen.

Dr. Adams consulted with pediatric surgeon Roshni Dasgupta, MD, who performed a laparoscopic biopsy of the mass on the now 7-week-old child. The mass was identified as a proliferating and inoperable hemangioma. Because the hemangioma could not be removed surgically, Dr. Adams initiated treatment of both the facial and internal hemangiomas with the chemotherapy drug Vincristine.

The use of steroids is standard in treating hemangiomas, but knowing the patient faced complicated cardiac surgery and the fact that steroids can interfere with wound healing, Vincristine was chosen as the course of treatment. This regimen was initiated at Cincinnati Children’s and then the treatments were resumed at her home hospital.

When the patient returned to Cincinnati Children’s, Dr. Adams brought cardiologist Russel Hirsch, MD, and cardiothoracic surgeons Peter Manning, MD, and Pirooz Eghtesady, MD, onto the case for consultation for the complicated aortic arch reconstruction, made more complex by the patient’s unusual aortic anatomy which resulted in atypical intracranial blood supply issues.

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Richard G. Azizkhan, MD  
Surgeon-in-Chief; Lester W. Martin  
Chair of Pediatric Surgery

As an international advocate for children’s rights and health, Dr. Azizkhan has made significant clinical and basic research contributions in angiogenesis and vascular malformations, lung development, airway reconstruction, solid tumors, trauma and injury prevention. He also has helped Cincinnati Children’s develop a major international medical education exchange program with Bosnia and Croatia.

Denise M. Adams, MD  
Inpatient Clinical Director, Division of Hematology/Oncology; Medical Director, Comprehensive Hemangioma and Vascular Malformation Center  
Named one of the Best Doctors in America in 2008, Dr. Adams served as visiting professor at the Vascular Anomalies Center of Children’s Hospital Boston before joining Cincinnati Children’s. Prior to that appointment, she served as assistant professor at the University of Vermont and has also served as director of the Multidisciplinary Vascular Malformations Program at Duke University Medical Center.

Recent Publications


Results and Follow-Up

Repair of the coarctation, performed by Dr. Eghtesady, took place in April of 2007 and was successful.

The child has now been off Vincristine since January of 2008 and is doing well. The hemangioma on her spleen is monitored with ultrasound and the latest findings show that it has decreased in size.

She recently had the hemangioma removed from the left inner side of her lip and she has begun laser treatment on the visible telangiectasias on her face. Now back home with her family, the child, age 2, has hit every developmental milestone and is doing well.

Discussion and Lessons Learned

This case serves as an ideal example of how little is known about PHACE syndrome, making it difficult to give a standard prognosis when there are no standard protocols for these children. The ability to closely consult among all involved disciplines helps to ensure that the best decisions are made in such challenging situations.

Not knowing the true prognosis is difficult and stressful for both families and staff. Having received one prognosis from the original hospital and then a very different one at Cincinnati Children’s required invaluable interaction, communication and education with the parents. Such cooperation was necessary for the parents’ comfort and confidence in their decision-making for their child.

Patients with such complex conditions need to be followed long-term in a multidisciplinary center where the prognostic implications of such conditions can be studied and followed, and new standards of care discovered.

Complex Issues Require Multidisciplinary Care

Vascular anomalies can range from uncomplicated vascular lesions to much more serious vascular tumors and malformations. The more serious and complicated vascular anomalies require multiple specialists for optimum care.

The Cincinnati Children’s Hemangioma and Vascular Malformation Center offers multidisciplinary care and expert assessment for patients with this complex and wide-ranging group of conditions.