Aortic arch repair in children with PHACE syndrome

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ABSTRACT

Background: PHACE syndrome is characterized by infantile hemangioma and developmental abnormalities of the brain, arteries of head and neck, and aortic arch.

Methods: We retrospectively reviewed The PHACE Syndrome International Clinical Registry to identify children with PHACE who had operative repair of aortic arch obstruction at Children's Hospital of Wisconsin.

Results: Seven patients (median 11 months, range 1 week-6 years) with PHACE required aortic arch reconstruction from 1996 to 2015. All needed complex surgical approaches (4 conduit grafts, 2 patch aortoplasties, 1 subclavian flap) to relieve the obstruction because of long-segment transverse and proximal descending aortic arch dysplasia that included multiple areas of stricture with adjacent aneurysmal dilatation. Aberrant origin of a subclavian artery was found in 6 of 7. The 3 children who had surgery after age 1 showed significant progression of the arch obstruction and/or adjacent aneurysmal segment dilatation after their initial infant evaluation. No deaths or perioperative complications occurred despite associated cerebrovascular arterial dysplasia in 5 of 7. Recurrent arch obstruction developed in 3 of 7 at an intermediate follow-up interval of 6.2 years (2 had interposition graft replacement at 8 and 11 years due to somatic growth; 1 had repeat patch aortoplasty 11 months after initial repair secondary to recurrent stenosis).

Conclusions: Extensive aortic arch reconstruction is commonly required in children with PHACE syndrome and coarctation due to the bizarre nature of the obstruction. Complete preoperative imaging is needed to fully characterize the aortic and cerebrovascular arterial anomalies. Recurrent obstruction is common given the non-native tissue techniques needed to relieve the arch anomaly. (J Thorac Cardiovasc Surg 2016;152:709-17)

The PHACE acronym was coined in 1996 to describe the association among malformations of the *P*osterior fossa, *H*emangiomas of the head and neck, *A*rterial, *C*ardiovascular, and *E*ye anomalies, and ventral developmental defects.¹ Forty-one percent of patients with definite PHACE

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Funding for this project was provided by the National Institutes of Health (Grant: 5 R01 AR064258-02).

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Received for publication Jan 26, 2016; revisions received March 21, 2016; accepted for publication March 27, 2016; available ahead of print May 7, 2016.

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0022-5223/\$36.00

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<http://dx.doi.org/10.1016/j.jtcvs.2016.03.082>

PHACE coarctation with concurrent great vessel malformations has increased ischemic risk.

Central Message

PHACE syndrome is associated with unusual aortic arch obstructions, requiring complex surgical solutions for repair.

Perspective

Thirty percent of children with PHACE syndrome have aortic arch obstruction and it is frequently unusual and long-segment with concurrent brachiocephalic and cerebrovascular arterial malformations. Extensive arch reconstruction using non-native tissue techniques is commonly required, and complete preoperative imaging is needed to fully characterize the aortic and cerebrovascular arterial anomalies.

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syndrome have intracardiac, aortic arch, or brachiocephalic arterial anomalies.^{2,3} The aortic arch anomalies observed in PHACE syndrome are especially complex and often involve long segments of the transverse aorta arch.¹⁻⁶ Approximately one-third of those will require surgical intervention.^{2,3} Most patients with PHACE syndrome also have anomalies of the brachiocephalic, cervical, and cerebral arteries.⁷ This combination of aortic arch, brachiocephalic, and cerebral vessel malformations leads to increased risk of ischemia and stroke, presenting a unique challenge to the surgical team.^{4,8} The surgical repair,

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Abbreviations and Acronyms

DHCA	= deep hypothermic cardiac arrest
MRI	= magnetic resonance imaging
MRA	= magnetic resonance angiography
NIRS	= near-infrared spectroscopy
PHACE	= P osterior fossa, H emangiomas of the head and neck, A rterial, C ardiovascular, and E ye anomalies, and ventral developmental defects

anesthesia, and perioperative care of these patients have not been previously reported. We describe our surgical experience with children who have PHACE syndrome and complex aortic arch and brachiocephalic anomalies requiring repair at Children's Hospital of Wisconsin.

METHODS

A total of 248 patients are enrolled and consented in the institutional review board–approved PHACE Syndrome International Clinical Registry and Genetic Repository at the Medical College of Wisconsin. The incidence of congenital cardiac disease in the registry has previously been reported; however, details regarding the surgical approach to repair of arch anomalies have not been fully described.³ Seven patients were identified for a detailed retrospective review because of a history of aortic arch abnormalities requiring surgical intervention performed at our institution. Three patients were previously reported by our group in 2008, but the details of the surgical approach were not published.² All relevant data, including clinical and cardiovascular/cerebrovascular imaging information, were reviewed by our multidisciplinary team.

RESULTS

Seven patients (median age 11 months, range 1 week–6 years) with PHACE required extensive aortic arch reconstruction from 1996 to 2015 (Table 1). Six of the 7 patients were girls, which is consistent with the well-described female predilection observed in PHACE syndrome. All needed complex surgical approaches (3 interposition grafts, 1 extra anatomic bypass graft, 2 patch aortoplasties, 1 subclavian flap) because of extensive arch dysplasia that included long-segment areas of stricture with adjacent aneurysmal dilatation to relieve the obstruction (Figures 1–7). Aberrant origin of a subclavian artery was found in 6 of 7, so that clinical assessment of the gradient by blood pressure measurement was impossible (all arm and leg arteries arose distal to the arch obstruction). None had associated intracardiac pathology, with anatomically normal aortic and mitral valves in all cases. Four patients presented with obstruction in the first month of life. The other 3 children who had surgery after age 1 year showed significant progression of the arch obstruction and/or aneurysmal segment dilatation after their initial infant evaluation. No

deaths or perioperative complications occurred despite associated cerebrovascular arterial dysplasia in 5 of the 7. One patient with severe bilateral cervical and cerebral arterial stenosis and an incomplete Circle of Willis had an acute ischemic stroke of left posterior temporal and occipital region 6 weeks after surgery that appeared unrelated to the procedure. This patient was included in a case series of children with acute ischemic stroke published in 2006.⁸ Recurrent arch obstruction developed in 3 of the 7 at intermediate follow-up (2 had conduit graft replacement at 8 and 11 years due to somatic growth; 1 had repeat patch aortoplasty for recurrent stenosis 11 months after an initial repair performed at an outside institution).

Of these patients, all underwent echocardiography as the initial imaging study to determine the intracardiac and aortic arch anatomy; 2 of the 7 patients underwent additional diagnostic imaging by cardiac catheterization to better assess the anatomy/perform direct pressure measurements. Magnetic resonance imaging/magnetic resonance angiography (MRI/MRA) of the aortic arch, brachiocephalic arteries, and cerebrovascular bed was performed in 6 of the 7 cases, with only the initial case in 1996 not undergoing that study, as the risk for brachiocephalic and cerebrovascular abnormalities was not recognized at that time. Intraoperative monitoring varied among patients. All patients had invasive arterial blood pressure monitoring, whereas central venous pressure monitoring was used in every procedure aside from the subclavian flap. Neuromonitoring techniques included near-infrared spectroscopy (NIRS), which was used to monitor cerebral perfusion in 8 procedures. Bilateral cerebral NIRS monitoring was used in 5 procedures and unilateral cerebral NIRS in 3 procedures. Somatic renal NIRS monitoring was used to assess perfusion distal to the aortic arch in 8 procedures. Including repeat repairs, cardiopulmonary bypass was used in 4 of 9 surgeries (3 of 3 repeat repairs). In addition, deep hypothermic cardiac arrest (DHCA) with antegrade cerebral perfusion was used in 2 of 9 operations.

COMMENT

Infantile hemangiomas are common, and approximately 30% of newborns with large hemangiomas of the head meet the diagnostic criteria for PHACE.⁷ The infantile hemangiomas associated with PHACE have a characteristic, segmental distribution usually involving large segments of the face, neck, and scalp (Figure 8). There has been increased recognition that patients with features of PHACE may lack the facial hemangioma, but have large segmental infantile hemangioma located on the posterior scalp/neck and the upper chest (as seen in case 5). Arterial anomalies are the most frequent noncutaneous finding in PHACE and appear to preferentially affect the aorta and

TABLE 1. Summary of cases

Case ID	Age at presentation	Clinical presentation	Arch and vascular anomalies	Age of surgery	Repair description	Follow-up repair (age)	Current status
1	3 wk	<ul style="list-style-type: none"> Large bilateral forehead and scalp hemangioma Coarctation 	<ul style="list-style-type: none"> Left aortic arch Aneurysmal transverse arch dilation with juxtaductal stenosis Narrow and dysplastic left internal carotid artery and a dysplastic anterior communicating artery 	6 wk	<ul style="list-style-type: none"> Reverse subclavian flap 	None	No additional intervention with clinical and echocardiographic evidence of a widely patent aortic arch and no residual gradient during 17 y of FU evaluation.
2	6 wk	<ul style="list-style-type: none"> Left facial, scalp and neck hemangioma 	<ul style="list-style-type: none"> Left aortic arch Interrupted aortic arch Saccular aneurysm of the transverse arch Aberrant right SCA Narrowed left ICA, right MCA, and left vertebral artery 	2 mo	<ul style="list-style-type: none"> Interposition graft (10 mm) 	<ul style="list-style-type: none"> Extra-anatomic bypass graft (CPB) (11 y) 	No further evidence of significant gradient during 16 y of FU.
3	1 d	<ul style="list-style-type: none"> Posterior fossa cyst Large left facial hemangioma Heart murmur 	<ul style="list-style-type: none"> Left aortic arch Mid and distal arch stenosis with focal aneurysmal dilation Aberrant right SCA Diminished basilar artery Stenosis in left CCA 	5 d	<ul style="list-style-type: none"> Interposition graft (8 mm) 	<ul style="list-style-type: none"> Extra-anatomic bypass graft (20 mm) (CPB) (8 y) 	No evidence of residual gradient during 10 y of FU.
4	5 wk	<ul style="list-style-type: none"> Face, right orbit, scalp and neck hemangiomas 	<ul style="list-style-type: none"> Right aortic arch Long segment distal arch stenosis Distal arch aneurysm Aberrant left SCA/vascular ring Early bifurcation of the right CCA Long segment narrowing of the right ICA 	28 mo	<ul style="list-style-type: none"> Extra anatomic bypass graft (10 mm) 	None	<ul style="list-style-type: none"> Mild proximal conduit narrowing (25 mm Hg Doppler gradient) MRI evidence of significant regression of the transverse arch aneurysm at 4 y of age (2 y following surgery).
5	Neonate	<ul style="list-style-type: none"> Coarctation Large back and neck hemangioma-no facial hemangioma 	<ul style="list-style-type: none"> Left aortic arch Long segment distal arch stenosis 	11 mo	<ul style="list-style-type: none"> Cormatrix patch aortoplasty (outside institution) 	<ul style="list-style-type: none"> Patch aortoplasty (11 mo) 	<ul style="list-style-type: none"> No evidence of residual gradient during 3 y of FU. No further surgical intervention.
6	6 y	<ul style="list-style-type: none"> Large segmented hemangioma on left face and scalp 	<ul style="list-style-type: none"> Left aortic arch Transverse arch aneurysm Distal arch stenosis Aberrant right ICA and SCA Narrow left ICA and SCA Absent left vertebral artery 	6 y	<ul style="list-style-type: none"> Interposition graft (16 mm) (CPB) (DHCA) 	None	<ul style="list-style-type: none"> No evidence of residual gradient during 8 y of FU. No further surgical intervention.
7	8 mo	<ul style="list-style-type: none"> Large right face and scalp hemangioma Right cerebellar hypoplasia with cleft 	<ul style="list-style-type: none"> Right aortic arch Long segment distal arch stenosis Aberrant left SCA with vascular ring Dysplastic right ICA 	17 mo	<ul style="list-style-type: none"> Homograft patch angioplasty (CPB) (DHCA) 	None	<ul style="list-style-type: none"> No evidence of residual gradient during 2.5 y of FU. No further surgical intervention.

FU, Follow-up; SCA, subclavian artery; ICA, internal carotid artery; MCA, medial carotid artery; CPB, cardiopulmonary bypass; CCA, common carotid artery; MRI, magnetic resonance imaging; DHCA, deep hypothermic cardiac arrest.

FIGURE 1. Left aortic arch with normal brachiocephalic origins and mild transverse arch aneurysmal dilatation (AN) immediately distal to the left subclavian artery (LSC) origin with severe narrowing (arrow) into the proximal descending thoracic aorta (A) and normal intracardiac anatomy. The patient underwent surgical coarctation repair at 6 weeks of age using a reverse subclavian flap technique (B).

medium-sized arterial vessels of the chest, neck, and head.^{3,5,6,8-13} In a 2013 analysis of the International PHACE Syndrome Registry the 62 (41%) of 150 had cardiac, aortic arch, or brachiocephalic anomalies. Coarctation was present in 28 (19%) of the 150, and 23 (15%) of 150 required surgical intervention of arch obstruction.³ This report emphasizes the complex nature of the aortic anomalies, and the need for complex surgical strategies for aortic arch reconstruction. The arch

obstruction is most often long-segment, rather than the typical discrete juxtaductal narrowing seen in coarctations of patients without PHACE syndrome. The obstruction is also frequently characterized by regions of arch narrowing or interruption with adjacent segments of marked aneurysmal dilatation that require resection. In addition, the obstruction may be difficult to appreciate clinically in those with PHACE syndrome because of the commonly associated aberrant subclavian origin (so that both

FIGURE 2. Markedly aneurysmal transverse aorta with poorly visualized continuity to the descending aorta by echocardiography with normal intracardiac anatomy. Aortic angiography demonstrated a left aortic arch with interruption immediately distal to a large (25-mm diameter) transverse arch aneurysm (AN) with the descending thoracic aorta supplied by the ductus arteriosus (A). Both the left (LSC) and right (RSC) subclavian arteries arose from the descending aorta with a common carotid giving rise to both the right (RC) and left (LC) carotid arteries. The patient underwent surgical aortic arch reconstruction with placement of a 10-mm diameter interposition graft (B). Her postoperative recovery was uneventful, but she had a stroke at 3 months related to her cerebrovascular anomalies. The interposition graft became progressively obstructive related to somatic growth, requiring placement of an extra-anatomic bypass graft using cardiopulmonary bypass (CPB) at age 11.

FIGURE 3. Echocardiography identified coarctation with unusual areas of dilatation and narrowing in the transverse aorta with normal intracardiac anatomy. Aortic arch magnetic resonance imaging/magnetic resonance angiography demonstrated a left aortic arch with multiple areas of aneurysmal (AN) dilatation with strictures (arrows) between the aneurysms and into the descending thoracic aorta (A). Both the left (LSC) and right (RSC) subclavian arteries arose from the descending aorta with a common carotid giving rise to both the right (RC) and left (LC) carotid arteries. The patient underwent surgical arch reconstruction with placement of an 8-mm diameter interposition graft (B). The interposition graft became progressively obstructive related to somatic growth, requiring placement of an extra-anatomic bypass graft using cardiopulmonary bypass (CPB) at age 8.

subclavian arteries arise distal to the obstruction, making 4 extremity blood pressure uninformative in identifying the arch gradient).

Our single-center surgical experience with PHACE-associated arch obstruction describes the extensive aortic arch reconstruction required to relieve the obstruction and remove aneurysmal segments. This is in contrast to the surgical approach needed for “typical” discrete coarctation found in patients without PHACE. Several recent reviews of surgical techniques used in repair of isolated coarctation describe the extended end-to-end anastomosis (with resection of the narrowed segment and direct re-anastomosis of the normally developed proximal and distal arch segments adjacent to coarctation) as the procedure used in more than 90% of cases.¹⁴⁻¹⁷ It is the technique of choice at our institution because it involves only native tissue, without placement of foreign material grafts or patches. In this series, however, none of the 7 patients could have an extended end-to-end repair because of the long-segment involvement of the aorta. Most (6/7) required non-native tissue reconstruction techniques (interposition graft in 3, bypass graft in 1, and patch aortoplasty in 2) and the other case used subclavian flap angioplasty. A contemporary review of coarctation techniques from Texas Children’s Hospital found that only 2 of 343 children required an interposition graft from

1995 to 2013.¹⁷ The use of more extensive reconstructive techniques for coarctation is avoided because it increases the risk of later reintervention. Reintervention rates for children after isolated coarctation repair are <5% at intermediate follow-up in recent series.^{6,15,17} This is in contrast to our experience, with 3 of the 7 children requiring reoperation for their arch anomaly.

Of additional perioperative concern are the frequent (90%) abnormalities observed in the arteries of the head and neck observed in patients with PHACE syndrome. These abnormalities are distinctive, analogous to the long-segment narrowing and dysplasia observed in the aorta.¹⁰ These developmental arterial anomalies have occasionally resulted in acute ischemic stroke in children with PHACE.^{4,8} Therefore, it is imperative that preoperative evaluation includes MRI/MRA imaging of the head and neck vasculature. Six of 7 of our patients had abnormalities of the medium-sized arteries of the neck with 3 of the 7 at severe risk for watershed ischemia given narrowing of the ipsilateral carotid artery and lack of collateral flow. Standard intraoperative monitoring usually includes electrocardiogram, pulse oximetry, capnography, noninvasive blood pressure, central venous pressure, and arterial (right radial and femoral) blood pressure. In PHACE cases, advanced neurophysiologic monitoring is indicated given the increased risk for

FIGURE 4. Newborn screening echocardiography revealed a right aortic arch with mild turbulence at the distal arch but no significant gradient and normal intracardiac anatomy. A heart murmur was noted during her 2-year old well-child examination, and repeat echocardiogram showed progressive aortic arch obstruction. Cardiac magnetic resonance imaging/magnetic resonance angiography and cardiac catheterization confirmed a right aortic arch with a common carotid giving rise to both the right (RC) and left (LC) carotid arteries and aberrant origin of the left subclavian artery (LSC) from the Kommerell diverticulum, forming a vascular ring. There was long segment mid and distal transverse arch hypoplasia with significant discrete narrowing just proximal to the origin of the right subclavian artery (RSC) and a large saccular aneurysm immediately distal to arch narrowing (A). There was a large intrathoracic hemangioma intimately adjacent to and surrounding the aneurysm. There was long segment mild hypoplasia of the aortic isthmal segment to the descending thoracic aorta immediately distal to the saccular aneurysm (arrows). To avoid the need for cardiopulmonary bypass, a 10-mm diameter extra-anatomic conduit was placed from the ascending to descending aorta (B) using 2 surgical approaches: a left thoracotomy approach to perform the distal anastomosis for the conduit (as well as vascular ring division), and a median sternotomy to perform the proximal anastomosis of the conduit. This was successful, as bypass was not needed; the aortic aneurysm was not resected because of concerns that it could not be removed without trauma to the adjacent hemangioma. Her postoperative course was complicated by prolonged chylous drainage, necessitating thoracic duct ligation, but she has otherwise done well. AN, Aneurysm.

ischemic brain injury and requirement for cardiopulmonary bypass.¹⁸⁻²⁰ Cerebral NIRS monitoring provides the continuous noninvasive assessment of cerebral regional oxygen saturation (RSO₂C), which reflects both regional cerebral venous saturation and cerebral oxygen delivery.²¹ Bilateral RSO₂C monitoring has been shown to be effective in detection of differential hemispheric perfusion in infants undergoing aortic arch reconstruction and is especially useful in PHACE syndrome as arterial anomalies are usually unilateral.¹⁹⁻²⁸ Somatic renal NIRS monitoring should be used in all patients with coarctation to assess distal perfusion.

Although not seen in our cohort described here, children with PHACE syndrome are also at increased risk for respiratory complications secondary to airway

obstruction.²² Subglottic and mediastinal hemangiomas increase the risk for airway complications in these children.²³ All patients with PHACE receive preoperative screening MRI/MRA of the head and neck, and our radiologist specifically evaluates the airway. If there is enhancement in the subglottic region, patients are referred to otolaryngology for bronchoscopy. External compression also may occur secondary to vascular ring formation with right-sided aortic arch anomalies.⁷

CONCLUSIONS

Extensive aortic arch reconstruction is commonly required in children with PHACE syndrome and coarctation due to the unusual and long-segment nature of the obstruction. Complete preoperative imaging is needed to

FIGURE 5. Arch magnetic resonance imaging/magnetic resonance angiography (MRI/MRA) showed a left aortic arch with long segment aortic atresia (arrows) extending for 1.5 cm from the aortic isthmus to the descending aorta (A). The transverse arch and brachiocephalic origins were normal. The patient underwent coarctation repair at an outside institution via a left thoracotomy with Cormatrix patch aortoplasty (B). She presented at 6 months old to our institution because of a large hemangioma on the back. Neck MRI/MRA demonstrated a left paraspinal and intraspinal (C7 to T9) hemangioma adjacent to the arch obstruction. Echocardiography revealed re-coarctation with long-segment recurrent narrowing in the descending thoracic aorta with otherwise normal intracardiac anatomy. She developed progressive upper extremity hypertension and underwent reoperation with homograft patch aortoplasty using homograft tissue at 11 months. *Ao*, Aorta.

fully characterize the aortic, brachiocephalic, and cerebrovascular arterial anomalies. Late and progressive stenosis and/or significant aneurysm formation can develop

after 1 year. Recurrent obstruction is common given the non-native tissue techniques needed to relieve the arch anomaly.

FIGURE 6. Echocardiography at 4 weeks of age showed a left aortic arch and a mildly hypoplastic distal transverse arch with a gradient of 20 mm Hg with normal intracardiac anatomy. The patient was lost to cardiology until 6 years of age, when echocardiography identified a left aortic arch with a moderate-sized saccular aneurysm (AN) in the mid transverse aortic arch between the left common carotid (LC) and left subclavian arteries (LSC) that was not appreciated at the neonatal study (A). There was aberrant origin of the right subclavian artery (RSC) from the descending thoracic aorta. The previously appreciated transverse arch narrowing immediately distal to the aneurysm appeared more significantly narrowed. Because of the unusual anatomy, she underwent cardiac catheterization to assess both the size of the aneurysm and the peak-to-peak arch obstruction. The gradient across the obstruction was mild at 20 mm Hg, but the size of the aneurysm was felt to warrant resection with aortic arch reconstruction. Elective surgical aortic arch reconstruction was performed using 16-mm diameter Hemashield interposition graft (B). *RC*, Right carotid artery; *Ao*, aorta.

FIGURE 7. Self-referred for a second opinion and echocardiography showed a right aortic arch with distal arch obstruction and normal intracardiac anatomy. Cardiac magnetic resonance imaging confirmed a right aortic arch with aberrant origin of the left subclavian artery (LSC) from a Kommerell diverticulum that completed a vascular ring (A). There was moderate long-segment transverse arch hypoplasia (arrows) immediately distal to the origin of the right subclavian artery (RSC) extending to the descending thoracic aorta at the origin of the diverticulum. She underwent elective arch reconstruction with vascular ring division at 17 months. The distal transverse arch hypoplasia required homograft patch angioplasty because of the length of the obstruction, and the ring was divided without manipulation of the diverticulum (B). RC, Right carotid artery; LC, left carotid artery.

Conflict of Interest Statement

Beth Drolet reports grant support from Pierre Fabre. Michael Mitchell reports equity in Ariesa Diagnostic, TAI Diagnostic, and MD Interactive. All other authors

have nothing to disclose with regard to commercial support.

The authors acknowledge Shawna Brown for the work in maintaining the International PHACE Syndrome Registry.

FIGURE 8. Case 4 with large segmental hemangioma of the right side of the face, involving the orbit and extending across the posterior scalp to the left parotid region.

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Key Words: congenital cardiac, aortic arch, subclavian artery, pediatric perioperative management