CASE REPORTS

Aortic aneurysm resulting from umbilical artery catheterization: Case report, literature review, and management algorithm

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Aortic aneurysms in infants and children are quite rare. The use of umbilical artery catheters in the management of critically ill neonates has been associated with infection and subsequent aneurysm formation. There have been 46 cases reported (including our own); most of the aneurysms have been located in the abdominal aorta and have displayed saccular morphology. Through an analysis of the literature, we identified two factors that had significant correlation with improved survival: diagnosis before surgery and surgical repair ($P < .05$). This report presents the case of a 23-day-old neonate with an abdominal aortic aneurysm and hypertension. On the basis of our literature review, we offer a management algorithm for this rare but very serious complication of umbilical artery catheterization. (J Vasc Surg 2001;33:419-24.)

Over the past 30 years, invasive monitoring has become an integral aspect of the management of critically sick neonates. Although the additional information accrued through the use of umbilical artery catheters is often very helpful and necessary, this form of invasive monitoring has potential significant complications; infection, hypoglycemia, bladder rupture, paraplegia, and umbilical artery rupture have all been described.[1-8] In addition, coarctation, dissection, thrombosis, and aneurysmal degeneration of the aorta and its major branches have also been reported.[1-4,9]

Although umbilical artery catheterization was first reported in 1962,[10] its association with subsequent aortic aneurysmal degeneration was not described until 1970.[11] Since then, more than 35 cases of aortic aneurysm resulting from umbilical artery catheters have been reported; however, a consistent method of evaluation and management has not evolved.[1-4,12-20] The purposes of this article are to (1) provide an overview regarding the causes of aortic aneurysms in children, (2) report another aortic aneurysm resulting from umbilical artery catheterization, and (3) suggest a diagnosis and treatment algorithm for this complication.

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Competition of interest: nil.

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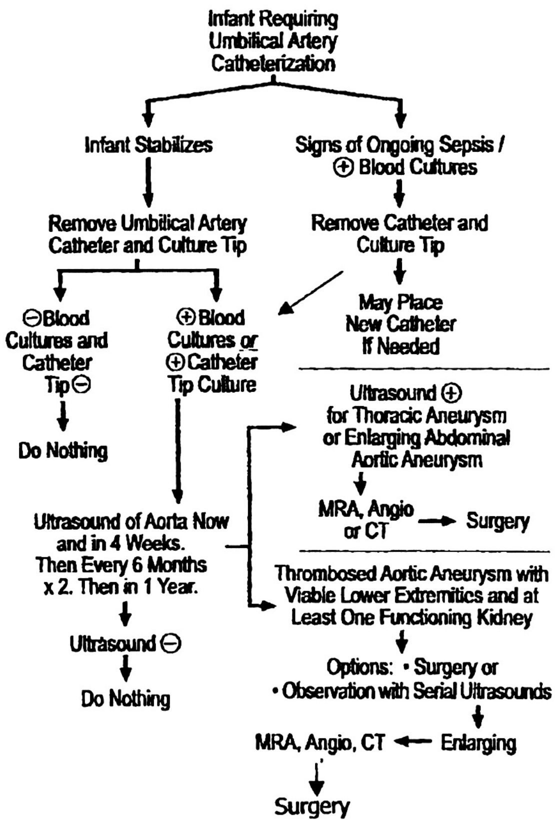
METHODS

Data acquisition for this analysis involved an extensive review of the literature, translation of foreign case reports, telephone calls to get updated information on previously reported cases, and review of our own recent experience. The data acquisition focused on aneurysm morphology and location, associated infections, method of diagnosis, timing of diagnosis, treatment, and survival. Statistical univariate analysis of survival relative to treatment modality and timing of diagnosis was made by means of the Fisher exact test. A multiple logistic analysis was performed to further assess factors that correlate with survival; it combined preoperative diagnosis and surgery. Significant differences were defined as those with P values of less than .05. Finally, through use of the data, a diagnosis and management algorithm was developed.

CASE REPORT

The patient was the female child of a 17-year-old mother (gravida 0, para 0). The infant was delivered by cesarean birth because of breach position; the birth weight was 2500 g. The pregnancy was significant for absence of prenatal care, oligohydramnios, and group B *Streptococcus*-positive cultures. The patient had respiratory distress at birth and required supplemental oxygen, with subsequent intubation on day 2 of life, because of progressive tachypnea. The patient remained on the ventilator for 6 days; use of a high-frequency oscillating ventilator was required. No surfactant was given. The patient was given two doses of dexamethasone before extubation.

Her hospital course during the first 3 weeks of life was significant for pneumonia, anemia requiring transfusion, hyperbilirubinemia, and the need for hyperalimentation. On days 7, 10, and 11 of life, the patient had blood cultures positive for methicillin-



Diagnostic and treatment algorithm for aortic aneurysm resulting from umbilical artery catheterization.

resistant staffers; this was treated with intravenous vancomycin, gentamicin, and rifampin. The umbilical artery catheter was discontinued on day 7 of life.

Beginning on day 11 of life, the patient was noted to have hypertension refractory to aggressive medical management, which included the use of hydralazine, lasix, and captopril. Her workup included an echocardiogram, the result of which was normal, and an abdominal ultrasound scan, which revealed an abdominal aortic aneurysm. This was treated conservatively. Because of worsening hypertension and the increasing size of the aneurysm on serial ultrasound scans, the patient was transferred to our institution on day 23 of life. A nipride drip was required to control her hypertension. A repeat ultrasound scan and a computed tomography (CT) scan revealed an enlargement of the abdominal area aneurysm.

The patient subsequently underwent surgical repair of the aneurysm through use of a Gore-Tex graft (W. L. Gore & Associates, Elkton, Md). The aneurysm extended from just distal to the superior mesenteric artery to the bifurcation of the aorta, with extension into the left proximal iliac artery. The aneurysm involved both renal vessels, which required reimplantation as a Carrel patch into the aortic graft. The patient required reexploration 24 hours postoperatively because of bleeding. The remainder of the postoperative course was significant for high-output renal failure resulting from acute tubular necrosis, which resolved.

Table I. Aneurysm location (N = 46)

Abdominal aorta	43% (n = 20
Thoracic aorta	30% (n = 14)
Iliac artery	15% (n = 7)
Thoracoabdominal	12% (n = 5)

The patient was able to be weaned off her antihypertensive medications immediately postoperatively, with no further episodes of hypertension. She was treated with 4 weeks of rifampin therapy as well as a total of 7 weeks of vancomycin therapy. She was subsequently discharged home on day of life 63 with a patent aortic graft and palpable femoral pulses bilaterally and normotensive.

RESULTS

Literature review. The data acquisition involved review of 26 articles—14 case reports, 4 review articles, and 8 papers pertaining to umbilical artery catheter complications other than aneurysm formation. In addition, six telephone contacts were made to update individual case reports.

Aneurysm location and morphology. Of 46 cases reported, aneurysms developed in the abdominal aorta in 20 (43%); 14 (30%) aneurysms occurred in the thoracic aorta, 7 (15%) occurred in the iliac artery, and 5 (12%) involved the thoracoabdominal aorta (Table I). As can be seen in Table II, most of the aneurysms (62%) were saccular; 28% were fusiform, and 10% were reported as pseudoaneurysms or of unknown morphology.

Associated infection. There was a very strong association between infection and subsequent aneurysm formation. A documented infection, defined as a positive blood culture, positive culture from the umbilical artery catheter, or positive intraoperative culture from the aneurysm wall, was found in 86% of the cases. *Staphylococcus aureus* and *Staphylococcus albus* were the most common microorganisms, being cultured in 79% of the cases (Table III).

Timing of diagnosis, treatment modality, and survival. Two factors were identified as significantly correlating with survival: timing of diagnosis and surgical repair. Specifically, those children diagnosed before laparotomy and those who underwent aneurysm repair had a statistically significant increase in probability of survival ($P < .05$). These results are reflected in Tables IV and V. When these two factors were analyzed together through use of multiple logistic regression analysis, both remained significant with a P value less than .01 (Table VI). This means that *both* factors significantly contributed to outcome, the statistically stronger element being surgical repair.

DISCUSSION

Aortic and iliac aneurysms are rare in children. There are several etiologies, and each can have a significant effect on the location of the aneurysm, the age of presentation, and the mode of treatment. With respect to cause, these aneurysms can be separated into four groups: congenital, inflammatory, infectious, and associated with iatrogenic trauma.

The *congenital* aneurysms cover the spectrum from

Table II. Aneurysm morphology

Saccular	62%
Fusiform	28%
Pseudoaneurysm or unknown	10%

Table III. Microorganisms cultured

Staphylococcus	79%
Klebsiella	7%
Pseudomonas	4%
Enterococcus	4%
Candida	3%
Enterobacter	3%

Organisms were present in 86% of cases. All microorganisms that were cultured are listed.

primary aneurysms seen at birth to aneurysms that developed subsequently as a result of congenital abnormalities, such as aortic coarctation, valvular stenosis, cystic medial necrosis, and Ehlers-Danlos and Marfan syndromes. These "secondary" aneurysms tend to be located in the thoracic aorta and are usually detected in children aged 4 to 15 years. The known exceptions to this observation are children born with tuberous sclerosis, who present with aneurysms at ages ranging from newborn to adolescence.

Inflammatory arteritides, such as Takayasu's disease, Kawasaki syndrome, polyarteritis nodosa, and granulomatous aortitis, may also cause aortic aneurysms in children. These aneurysms tend to be complications of the primary aortitis and are very rare.

The final two etiologies of childhood aortic aneurysms are *infection* and *iatrogenic trauma.* These are presented together because an association between umbilical artery catheterization and infection and subsequent aortic aneurysm has been noted.[11]

Although infection is responsible for only 2.5% of all aortic aneurysms, it is associated with 80% of the aneurysms caused by umbilical artery catheters. The exact mechanism leading to aneurysm formation is not clear. However, there appear to be two likely explanations.

First, under normal conditions, the intact intima is resistant to infection. Once the intima has been violated by trauma from the umbilical artery catheter tip, the vessel wall becomes susceptible to an infective source. In this scenario, the catheter not only causes intimal disruption but also harbors the infective microorganism. In most instances, the microorganism is *S aureus* or *S albus.* A concomitant factor in this process is the clinical state of the patient. Umbilical artery catheters are often placed in neonates who are hypoxic because of compromised cardiorespiratory function. Krist et al[21] have shown that prolonged hypoxemia damages the aortic wall and increases aortic susceptibility to infection.

The second explanation for mycotic aneurysm formation is obstruction of the aortic vasa vasorum with infective thrombus. Again, this thrombus is thought to originate from an infected catheter tip. Because the vasa vasorum supplies nutrients and oxygen to the aortic wall, a localized obstruction can lead to a compromised vessel that is susceptible to infection. In the setting of intimal damage and obstructed vasa vasorum, the infection causes a localized inflammatory process that weakens the vessel wall and leads to subsequent aneurysm development.[22] In our case, the pathology specimen demonstrated aortic rupture with numerous giant cells; there was no evidence of obstruction of the vasa vasorum. The exact mechanism of aortic wall infection and aneurysmal function could not be determined. In the cases published, we were not able to identify a specific pathophysiology of infection or arterial damage that resulted in aneurysm formation. The location of the catheter tip and the length of time that it is in place appear to be related to subsequent aneurysm formation. Drucker et al[3] reported the association between catheter tip location and site of subsequent aneurysm formation. Aneurysm formation appears to be more common if the catheter tip is in the thoracic aorta, whereas there is risk of hypoglycemia, visceral infarction, and aortic thrombosis when it is in the abdominal aorta. Symansky and Fox[23] observed that infants who had umbilical artery complications had a mean catheter duration twice that of infants who did not have complications.

Most children with aneurysms resulting from umbilical artery catheters present between the ages of 5 weeks and 5 years. The clinical presentation varies depending on the site of the aneurysm. A thoracic aortic aneurysm presents either with rupture or as an asymptomatic mass on a chest radiograph. Conversely, most aneurysms of the abdominal aorta present as hypertension or as asymptomatic abdominal masses, spontaneous rupture occurring in a minority.

Over the past three decades, heightened awareness regarding the potential sequelae of umbilical artery catheterization has increasingly allowed clinicians to make the diagnosis before rupture. Although 30 years ago most of these aneurysms in children were diagnosed at autopsy or laparotomy, most are now diagnosed preoperatively by means of ultrasound scanning, magnetic resonance imaging, CT, or aortography (Table VII).

Analyzing the reported cases, we identified two factors that significantly correlated with survival: timing of diagnosis and surgical repair. Specifically, those children diagnosed before laparotomy and those who underwent aneurysm repair had a statistically significant increase in probability of survival ($P < .05$). This indicates that the natural history of aortic aneurysms in children is similar to that in adults, despite the differing aneurysm etiologies.

Although it is intuitive that having a preoperative diagnosis and performing an elective repair of an aortic aneurysm should improve outcome, the statistical verification in this unique clinical setting has not been previously documented. Furthermore, the statistically significant survival benefit of preoperative diagnosis suggests the importance of clinicians having knowledge regarding the

Table IV. Effect of diagnostic timing on outcome

Timing of diagnosis vs outcome			
Timing	Outcome: dead	Outcome: alive	Total
Not diagnosed ahead	15	6	21
Diagnosed ahead	6	19	25
Totals	21	25	46
Tests			
Source	DF	–Log likelihood	R^2 (U)
Model	1	5.369978	.1693
Error	44	26.3406600	—
C total	45	31.710637	—
Total count	46	—	—
Test	χ^2	Probability > χ^2	
Likelihood ratio	10.740	.0010	
Pearson	10.348	.0013	
Fisher exact test	Probability		
Left	.998		
Right	.0016		
2-tail	.0026		
κ: 0.474286	SE: 0.130288		

κ measures degree of agreement.
*P < .0001.

Table V. Effect of treatment on outcome

Treatment vs outcome			
Treatment	Outcome: dead	Outcome: alive	Total
No surgery	17	4	21
Operated 9.05	4	21*	25
Totals	21	25	46
Tests			
Source	DF	Log likelihood	R^2 (U)
Model	1	10.49724	0.3309
Error	44	21.216914	—
C Total	45	31.216914	—
Total count	46	—	—
Test	χ^2	Probability > χ^2	
Likelihood ratio	20.987	< .0001	
Pearson	19.407	< .0001	
Fisher exact test	Probability		
Left	1.000		
Right	<.0001		
2-Tail	<.0001		
κ: 0.649524	SE: 0.112559		

κ measures degree of agreement.
*P < .0001.

association between umbilical artery catheterization and subsequent aortic aneurysm formation. In further support of the potential benefit of preoperative diagnosis, we found that of the four patients who had surgery and died, only one (25%) had a correct preoperative diagnosis. Conversely, of the 21 surgical patients who survived, 15 (71%) had correct preoperative diagnoses. Finally, as the incidence of preoperative diagnosis has increased over the last 30 years, the mortality has decreased from 58% before 1985 to 35% since 1985. This decline in mortality is likely the result of increased awareness of the disease process and improved imaging techniques, which lead to more elective surgical repairs.

The survival advantage in the surgically treated patients is very impressive. However, it may be equally instructive to focus on the medically treated group. Is there a good candidate for observation? On the basis of our review of the literature, we note that all of the patients who survived with nonoperative therapy had resolution of sepsis and thrombosis of their abdominal aortic aneurysms. This supports the concept that neonates and children may tolerate progressive aortic thrombosis. Even when the thrombosis extends to one kidney, blood pressure can be controlled with antihypertensive drugs, and nephrectomy, if needed, may be delayed until the child stabilizes. Conversely, surgical repair is indicated if the aor-

Table VI. Multivariate regression analysis: effect test

Source	*Nparm*	*DF Wald*	χ^2	*Probability* > χ^2
Diagnosed ahead	1	1	6.332779	.0119
Operated	1	1	11.011205	.0009

Table VII. Trends in timing of diagnosis

Timing of diagnosis	*1970-1984*	*1985-Present*
At laparotomy/autopsy	17	4
By preoperative imaging	9	16

Table VIII. Average age of patients

	Age (mo)	
Patient subset	*Alive*	*Dead*
Not diagnosed before laparotomy*	16.4	2.78
Diagnosed before laparotomy	14.9	0.79
No surgery	3.9	1.25
Surgery	17.5	6.3

*Patients not diagnosed before laparotomy were diagnosed at time of surgery or at autopsy.

toiliac aneurysm is causing uncontrolled sepsis, increasing rapidly in size, or causing cardiorespiratory deterioration.

It is important to point out the potential inherent bias with regard to survival after either operative or nonoperative management. Aneurysms in the surviving children were generally diagnosed at a later age (Table VIII). Therefore, the overall survival of these patients may in part be due to the fact that they had more stable aneurysms and less life-threatening disease processes.

It has been difficult to obtain good follow-up for determining long-term outcomes. The major complications are graft infection and graft stenosis. The latter can present with lower extremity ischemic symptoms and limb growth abnormalities. This makes it critical to have yearly follow-up until adulthood.

Cribari et al[1] suggest that because of their high propensity to rupture, all thoracic aneurysms be treated by surgical repair. The options for repair include resection and prosthetic replacement, resection with end-to-end anastomosis, and lateral resection and primary patch repair of the remaining aorta. The main advantage of the latter two options related to the exclusive use of autologous tissue, which is more resistant to recurrent infection and will allow for growth as the child develops. Obviously, the specifics of each clinical scenario will dictate which repair is the best option.

On the basis of this review, a diagnostic and treatment algorithm has been developed (Figure). It includes a high frequency of ultrasound scan surveillance in the first year, especially in the first 6 weeks. This approach reflects two aspects of our analysis. First, more than 50% of the aneurysms were diagnosed in children younger than 6 weeks, and more than 80% occurred in children during the first 2 years of life.[1-4] Second, there is a strong association between infection and subsequent aneurysm formation. Positive cultures were obtained in 86% of the cases. Therefore, identifying positive blood or umbilical artery catheter tip cultures helps select infants who may be at increased risk for subsequent aneurysm formation. It is imperative that these patients be followed with serial ultrasound scans. Third, the performance of angiography, CT, or magnetic resonance angiography after ultrasound scan diagnosis of a patent aneurysm is necessary to plan subsequent surgery.

The algorithm suggests that early diagnosis and surgical repair will most likely lead to a successful outcome. Observation may be appropriate in a child whose aneurysm is thrombosed and who has viable lower extremities. Finally, this algorithm, like all others, should be used as a guideline when applied to individual patients.

Although congenital and inflammatory disorders contribute to the development of aortoiliac aneurysms in children, those resulting from umbilical artery catheters and infection have predominated in most reports. Increased emphasis on prevention and diagnosis have markedly improved the survival of this dreaded complication. In most of the other recent cases, as in our own, diagnosis and treatment carried out in a timely fashion led to successful outcomes.

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