

Interventional Therapy for Coarctation of the Aorta

Transcatheter treatment of native and recurrent coarctation of the aorta can be a successful procedure at acute and intermediate follow-up.

BY DANIEL R. TURNER, MD, FAAP, FACC, AND THOMAS J. FORBES, MD, FACC, FSCAI

A 62-year-old man, with a 20-year history of treatment for essential hypertension with four antihypertensive medications, tripped while on a boat, bumped his head, and fell unconscious into the water. He was completely recovered upon his arrival to the emergency department, but due to the circumstances, a chest x-ray was obtained. Rib notching was noted, and further evaluation found severe, native coarctation of the aorta. He successfully underwent transcatheter placement of a Palmaz Genesis XD stent (Cordis Corporation, Bridgewater, NJ) to repair the coarctation, with resolution of his hypertension and discontinuation of all antihypertensive medications within 6 months of the procedure.

Coarctation of the aorta occurs in 0.04% of the population and accounts for approximately 5% of all congenital heart defects.¹

Coarctation is associated with a bicuspid aortic valve in 60% of the cases, although of more concern is that 10% have an association with a cerebral aneurysm.² Therefore, anyone presenting with coarctation of the

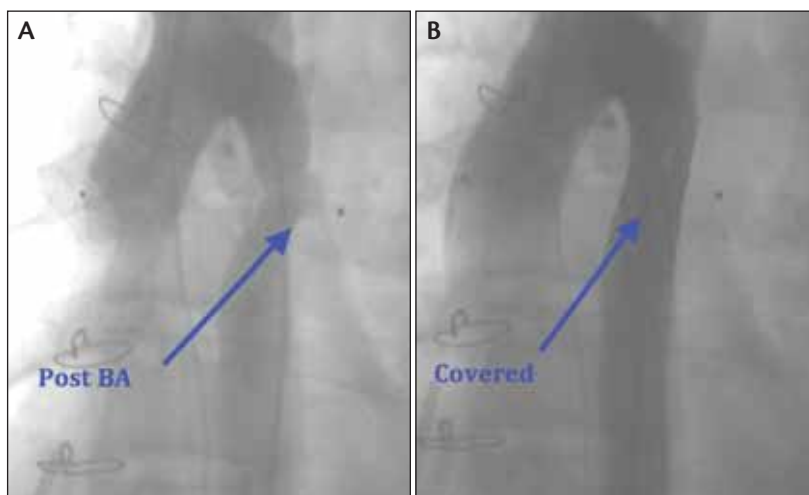


Figure 1. Status after balloon dilation of a native coarctation of the aorta in a 10-year-old girl (A). Three years later, she presented with reobstruction and late aneurysm formation. A covered CP Stent (NuMed, Inc., Hopkington, NY) was successfully used to treat both the reobstruction and the aneurysm (B).

aorta beyond early childhood should undergo further head imaging to rule out a berry aneurysm. Although the case presented is an extremely unusual presentation for native coarctation of the aorta, it does show what progress has been made in the treatment of coarctation of the aorta during the past 20 years. In fact, in patients older than 4 years or more than 15 kg in weight, transcatheter therapy for both native and especially recurrent coarctation of the aorta has become the standard of care in the majority of institutions participating in the Congenital Cardiovascular Interventional Study Consortium (CCISC). Recent recommendations from the American Heart Association/American College of Cardiology guidelines state that the coarctation of the aorta should be repaired in patients with > 20 mm Hg peak-to-peak gradient, or in patients with < 20 mm Hg gradient with significant angiographic/imaging evidence of a narrowing (level of evidence C). Controversy continues to surround the role of transcatheter treatment of native coarctation of the aorta, with more support being present for use of balloon angioplasty in the treatment of recurrent coarctation of the aorta.³ In this article, we discuss the present state of, and recent improvements in, transcatheter treatment of native and recurrent coarctation of the aorta in children and adults.

BALLOON ANGIOPLASTY

Balloon angioplasty was first described in 1983 by Lock et al.⁴ Since then, balloon angioplasty has gained widespread acceptance for the treatment of recurrent coarctation of the aorta in all age groups. Although balloon angioplasty of native coarctation in infants is quite controversial and is regarded by most to be a palliative procedure in high-risk patients,⁵ angioplasty of discrete, native coarctation in children and adults has recently shown excellent results. Walhout et al reported on 29 patients between 15 and 71 years of age with discrete, native coarctation of the aorta after successful balloon angioplasty procedures. No adverse events were encountered, with the peak systolic gradient decreasing from 52 mm Hg to 7.6 mm Hg immediately after balloon angioplasty. Intimal tears were detected in eight of 29 patients, with no evidence of acute dissection or aneurysm formation. In four of eight patients, there was resolution of the tear at 3-month angiographic follow-up. At a mean follow-up of 8.5 years, no dissection or late aneurysm formation was observed. One patient (3%) developed reobstruction, which required reintervention.⁶ Similar results were obtained by Hassan et al in 58 patients, with a mean follow-up of 13.4 years. In this study, 7% developed aneurysm and 8% had subop-

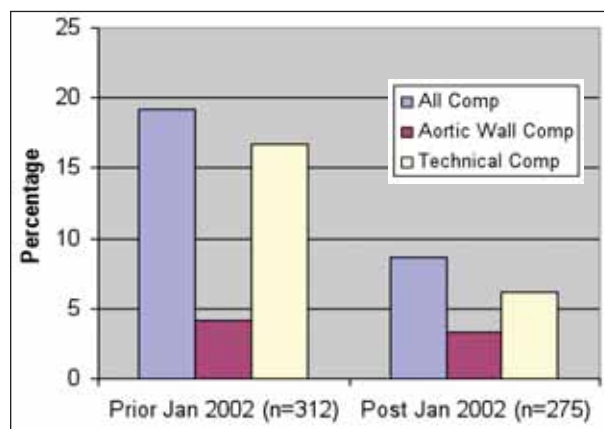


Figure 2. Complication trend over 14 years in stenting coarctation of the aorta.

timal outcome, with a peak gradient > 20 mm Hg being observed.⁷

The technique for performing balloon angioplasty for recurrent native coarctation of the aorta has evolved during the past 20 years. In infants, low-profile balloon angioplasty catheters up to 10 mm in diameter achieve sufficient atmospheres for adequate dilation of the coarctation segment. More frequently, larger-diameter, higher-profile balloon angioplasty catheters are required for adequate expansion of the narrowing. Balloon size is typically equal to or slightly larger than the diameter of the smallest segment of normal arch. Care must be taken to properly remove air from the balloon, as air emboli strokes have been reported to result after balloon rupture.

Aneurysm formation at intermediate and late-term follow-up does not appear to be related to the presence of an intimal tear at the time of the initial procedure. The majority of aneurysm formation appears to be small and discrete, requiring no therapy; occasionally, however, covered stent placement or surgical intervention is required for late aneurysm formation in patients after balloon dilation of native recurrent coarctation (Figure 1A, B).⁸

In summary, balloon angioplasty remains a controversial procedure in neonates and infants with native coarctation of the aorta. Satisfactory results may be achieved in older children and adults, with up to a 7% risk of aneurysms in a recent study.⁷ Balloon angioplasty is the treatment of choice for recurrent coarctation of the aorta in patients of all age groups. Intermediate and late aneurysm formation has been described, and integrated aortic arch imaging should be obtained in all patients within the first 5 years from initial intervention. The results of more than 90 patients undergoing bal-

loon angioplasty of native and recurrent coarctation of the aorta, with follow-up imaging 1 to 3 years after initial intervention, will be reported by the CCISC later this year.

STENT THERAPY FOR COARCTATION OF THE AORTA

Since the 1990s, stent treatment of native or recurrent coarctation of the aorta in children older than 4 years has become the treatment of choice at many institutions. In fact, at the 36 institutions participating in the Surgical versus Stent Treatment versus Balloon Angioplasty of Native or Recurrent Coarctation of the Aorta study, in children > 15 kg, stent placement is performed more than twice as commonly than balloon angioplasty and surgical treatment combined.

Stent placement for coarctation of the aorta was first performed in the early 1990s by O'Laughlin et al.⁹ The technique for intravascular stent treatment of coarctation of the aorta has evolved during the past 2 decades, which has led to a significant decrease in the complication rate (Figure 2).

OUTCOMES

Numerous studies have shown the safety and early efficacy of intravascular stent therapy, both acutely and at intermediate follow-up in the treatment of coarctation of the aorta.¹⁰⁻¹² In a multi-institutional retrospective study, intravascular stent placement was successful in 553 of 565 (98%) procedures. The definition of success was a systolic catheter gradient of ≤ 10 mm Hg after stent placement. The average peak systolic coarctation gradient acutely decreased from 31.6 to 2.7 mm Hg. The mean diameter of the coarctation segment increased from 7.4 to 14.3 mm.¹³

Complications in this study were divided into aortic wall and technical complications. Aortic wall complications were encountered in 22 of 565 patients (4%), including intimal tear, dissection, and aortic aneurysm. Aortic rupture was seen in three patients (each younger than 22 years) and in two-thirds of patients with recurrent coarctation of the aorta. One patient underwent successful placement of a covered stent, whereas the other two patients died 1 day and 6 months, respectively, later due to severe neurologic injury associated with

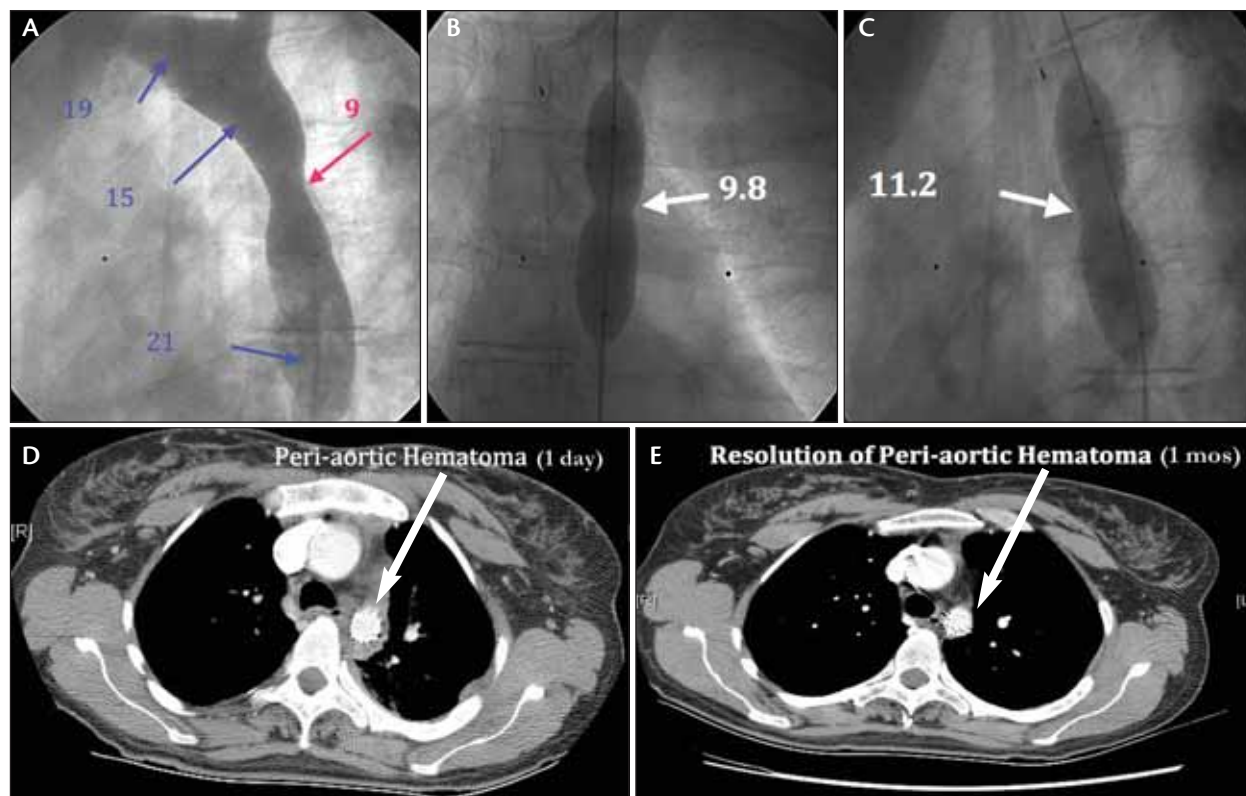


Figure 3. Native coarctation in a 43-year-old woman (A). Balloon compliance testing of the coarctation segment noted it to be noncompliant. A stent was placed across the coarctation segment and dilated to an 11- X 13.1-mm diameter (B and C). A follow-up CT scan obtained the next morning noted near transection of the thoracic aorta (D). A CT scan obtained 1 month later noted resolution of the periaortic hematoma (E).

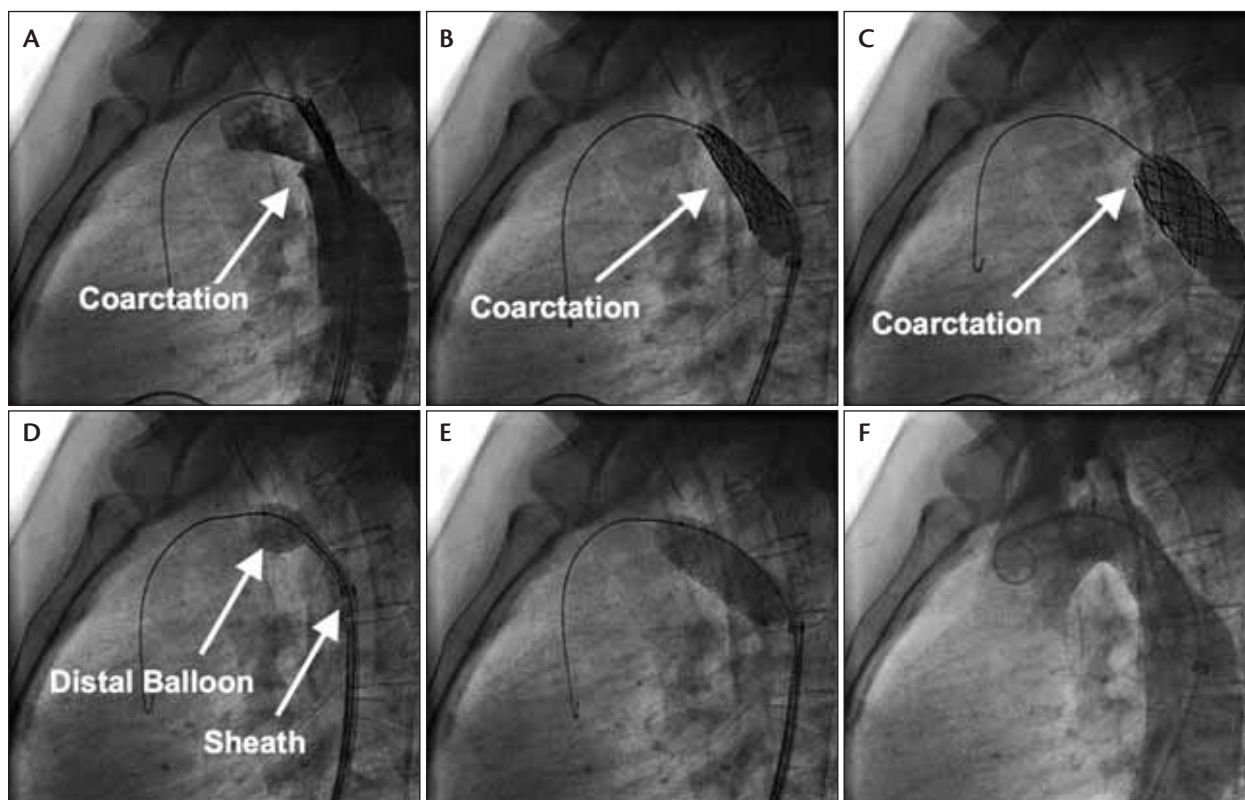


Figure 4. Etiology of stent migration with subsequent placement of a Palmaz Genesis XD stent (Cordis Corporation) across the coarctation segment. Figure A notes the CP Stent placed on balloon-on-balloon (BIB) catheter across the coarctation segment. Figures B and C note proximal-to-distal inflation of the outer BIB balloon. The waveform travels superiorly toward the coarctation segment, pushing the balloon/stent inferiorly, below the coarctation segment. Figures D and E show distal expansion of the balloon catheter with the Palmaz Genesis XD stent. The balloon catheter expands superiorly to inferiorly, preventing migration of the balloon/stent catheter inferiorly. Figure F depicts the stent properly positioned across the coarctation site.

the rupture. The rate of aortic wall complication remains relatively consistent before and after January 2002, remaining at 3.5% to 4% (Figure 2). Similar results have been observed in the current prospective CCISC experience. There is an increased likelihood of encountering acute aortic wall complications with (1) pre-stent balloon angioplasty, (2) location of the coarctation in the abdominal versus isthmus or transverse aortic arch, (3) age over 40 years, and (4) in patients' inherent vessel wall abnormalities.¹³⁻¹⁸ This is likely related to decreased aortic wall compliance and not to exceeding balloon-to-coarctation ratio standards ($> 3.5:1$). We personally encountered a near aortic rupture in a 43-year-old woman with a balloon-to-coarctation ratio $< 1.3:1$. A CT scan obtained the next day noted significant aortic wall injury. Complete resolution was noted 1 month later (Figure 3A through E).

Accurate assessment of aortic wall compliance is very difficult, with some interventionists advocating balloon

compliance testing. A compliant balloon, which equals the smallest dimension of the transverse aortic arch or descending aorta, is inflated between 2 to 3 atm. If a $> 30\%$ waist is seen in the balloon, the aorta is considered noncompliant, and the patient is likely at higher risk for standard bare-metal stent placement. Patients believed to be at high risk include adults over the age of 22 years and those with known connective tissue disorders (eg, Turner, Marfan, and Ehlers-Danlos syndromes). Although we do not routinely perform balloon compliance testing, a similar technique is used during initial stent deployment in patients thought to be at high risk. The balloon catheter and stent are gradually inflated, distally to proximally across the coarctation segment. Low inflation pressure is used. An "hour glass" deformity, in which the stent is not fully expanded across the coarctation segment, is frequently seen. As long as the stent is in a stable position, aggressive dilation of the stent during initial deployment is not performed.

Instead, a staged approach is used, with repeat dilation at 4 to 6 months. Although a perfect angiographic outcome is not the goal, repeat dilation is only performed if the patient remains hypertensive with an upper-to-lower blood pressure systolic gradient > 20 mm Hg.

It is difficult to accurately report the true prevalence of aortic wall injury at intermediate and long-term follow-up because of incomplete follow-up imaging after intravascular stent treatment of recurrent or native coarctation of the aorta. Only 27% (160 of 588) procedures were followed by integrated aortic arch imaging (magnetic resonance imaging/computed tomography [CT]/cardiac catheterization) in the CCISC coarctation study. Of the 160 patients, 16 underwent planned staged repeat dilation of the coarctation stent 6 to 14 months after the initial procedure. In the 144 remaining patients, 18 (12.5%) had evidence of aortic wall injury, including 13 aortic aneurysms. Four of the 13 patients required placement of a covered stent due to the large size of the aneurysm. Aortic wall injury at intermediate follow-up was associated with a balloon-to-coarctation ratio > 3.5 and prestent balloon angioplasty at the time of the initial procedure. Development of aortic wall injuries was not related to patient age or recurrent versus native coarctation of the aorta.¹⁹

The next most common abnormality at intermediate follow-up was in-stent restenosis secondary to neointimal proliferation within the stent ($n = 16$) or stent fracture ($n = 6$). In-stent restenosis at intermediate follow-up was associated with younger patient age, lower body weight, and smaller stent diameter at the initial procedure. Caution should be used when interpreting these data secondary to the small number of patients undergoing integrated aortic arch imaging at follow-up.¹⁹

TECHNICAL COMPLICATIONS

The greatest improvement in the overall complication rate for intravascular stent treatment of native or recurrent coarctation of the aorta has been in technical complications. Technical complications have decreased from $> 17\%$ before January 2002 to 5.3% after January 2002 (Figure 2). This trend continues to show gradual improvement in the current prospective CCISC study. The likelihood of encountering technical complications of any type is related to older patient age and the use of moderate sedation over general anesthesia.¹³ Stent migration is the most frequently encountered technical complication, occurring in 28 of 565 (5%) of procedures reported. The most common cause of stent migration is thought to be oversizing the balloon catheter relative to the transverse aortic arch, causing the balloon/stent to migrate distal to the coarctation segment during infla-

tion. This occurred in 14 of 28 of the stent migration procedures. The second most common cause of stent migration (nine of 28) was stent deployment using an undersized balloon. Clearly, balloon size is a critical decision, and many factors must be considered.

Newer techniques have been used to decrease cardiac output to stabilize the balloon/stent during delivery. Although adenosine or esmolol have been used in this manner,^{20,21} atrial or ventricular pacing offers a more reliable means to successfully decrease cardiac output.^{22,23} The heart is paced at a rate at which the systolic pressure decreases by 30% to 50% from baseline.

Usually, in older children and adults, this translates to a pacing rate of 160 to 185 bpm. Higher rates may be necessary for smaller children. No studies have proven that cardiac output reduction measures decrease the likelihood of stent migration.

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Use of the BIB catheter plays a theoretical role in decreasing the likelihood of stent migration, although retrospective analysis has shown this not to be the case.¹³ Alternate balloon inflation techniques may also be helpful. During initial inflation of the balloon, the sheath is pulled back to expose the stent but left covering the proximal shoulder of the balloon. During slow inflation, the balloon catheter fills distally to proximally, because the proximal part of the balloon cannot inflate. This allows delivery of the distal stent to the aortic wall before pulling the sheath back to deliver the more proximal stent across the coarctation segment. More accurate stent position may then be achieved. This technique and its potential advantage can be seen in a recent patient. In a 61-year-old woman, the transverse aortic arch measured 18 mm, and an 18-mm BIB balloon catheter/CP Stent (NuMed, Inc., Hopkington, NY) was advanced across the coarctation segment. The inner balloon had been previously inflated and properly positioned across the coarctation segment. Subsequent standard inflation of the outer balloon catheter noted proximal expansion of the balloon with subsequent balloon/stent migration to the midthoracic aorta (Figure 4). Subsequently, in the same patient, gradual inflation of the distal stent on a standard balloon catheter, as described previously, allowed eventual proper place-

ment of a second stent across the coarctation. Care must be used when performing this technique using covered/premounted stents, which may be at increased risk of stent slippage off the balloon catheter. This has not been observed with bare-metal stents that have been hand-crimped onto the balloon catheter.

NEW ADVANCES

The use of covered stents has gained widespread acceptance in other countries but remains unapproved in the United States. At this time, the Coarctation Stent Trial (COAST) is underway in the United States to evaluate the use of the CP Stent for the treatment of native and recurrent coarctation of the aorta in patients weighing less than 35 kg. Atrium Medical Corporation (Hudson, NH) is launching an international trial (winter 2009) using the Advanta V12 covered stent for treatment of coarctation of the aorta in children weighing greater than 30 kg. Preliminary reports indicate that covered stents may be redilated, at least in early follow-up, to adult size.^{24,25} Covered stents for treatment of balloon angioplasty aneurysms, complex coarctation of the aorta, and in patients with advanced age have been successfully used in 30 patients. No complications were encountered, with the stents remaining patent and in an excellent position on CT/magnetic resonance imaging performed 3 to 6 months later.²⁶ No fractures have been seen with the covered stents, which was observed in approximately 4% of the patients receiving the Palmaz Genesis XD stent at intermediate follow-up.¹⁹ The authors believe it is of paramount importance to have a covered stent available in the catheterization lab before imparting on bare-metal stenting of native or recurrent coarctation of the aorta, especially in the high-risk patient (ie, older adults). Although dissection and subsequent aortic rupture are thankfully rare, when they do occur, the only likely successful chance at salvaging the situation is emergent placement of a covered stent.

CONCLUSION

Transcatheter treatment of native and recurrent coarctation of the aorta is shown to be an acutely successful procedure at acute and intermediate follow-up. Significant improvements in avoiding technical complications have been observed during the past decade. Improved follow-up, both clinically and with integrated imaging, is required before we can determine if transcatheter treatment is superior to surgery. ■

Daniel R. Turner, MD, FAAP, FACC, is Associate Professor of Pediatrics at Wayne State University, Children's Hospital of Michigan, in Detroit. He has disclosed that he holds no

financial interest in any product or manufacturer mentioned herein.

Thomas J. Forbes, MD, FACC, FSCAI, is Associate Professor of Pediatrics at Wayne State University, and Director of the Cardiac Catheterization Laboratories, Children's Hospital of Michigan, in Detroit. He has disclosed that he holds no financial interest in any product or manufacturer mentioned herein. Dr. Forbes may be reached at (313) 745-5835; tforbes@dmc.org.

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