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ORIGINAL PAPER

Clinical characteristics of acute mitral regurgitation due to ruptured chordae tendineae in infancy—experience at a single institution

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Abstract In infants, acute mitral regurgitation resulting from ruptured chordae tendineae is very rare, but often fatal. There are a few case reports, but the characteristics and etiology of chordae tendineae rupture have not been elucidated. Our aim was to determine the clinical characteristics of idiopathic acute mitral regurgitation due to chordal rupture in infancy. A retrospective analysis was performed on ten consecutive patients, with a mean onset age of 4.6±1.3 months. Despite nonspecific initial symptoms, all patients developed respiratory distress and four required resuscitation within a few days (mean, 1.8±1.8 days). Chest radiographs showed pulmonary congestion with a normal or mildly increased cardiothoracic ratio in all ten patients. Laboratory data and electrocardiograms showed nonspecific findings. Echocardiography revealed ruptured chordae in all patients; locations were anterior (50%), posterior (20%), and both (30%). Surgical intervention was performed within 24 h of admission in eight patients (mean, 3.6±5.1 h). Pathological findings included inflammatory cells in six specimens and myxomatous degeneration in two. No bacteria were isolated from preoperative blood cultures, pathological tissues, or excised tissue cultures. Autoantibody levels were insignificant. Three preoperatively resuscitated patients developed neurological sequelae and arrhythmias occurred in four after mitral valve replacement. Acute onset and rapid deterioration in patients with ruptured chordae tendineae necessitates early surgical intervention to improve outcomes. Though the etiology remains unknown, onset is in infants approximately 4 months of age, suggesting a definite disease entity.

Keywords Acute mitral regurgitation · Ruptured chordae tendineae · Infant · Mitral valve replacement · Complication

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Introduction

Acute mitral regurgitation (MR) is characterized by acute onset, rapid deterioration, and failure of the left heart, frequently requiring surgical intervention [11]. MR results from various abnormalities, including leaflet, annulus, chordae tendineae, papillary muscle, and left ventricular (LV) wall lesions. In adults, acute MR often occurs in ischemic heart disease due to papillary muscle ischemia. However, in pediatric patients, acute MR is rare.

Rupture of the chordae tendineae of the mitral valve was first described by Corvisart in 1806 [5]. Thereafter, many cases were reported [14]. Reported etiologies include ischemia [1, 13, 14], infectious endocarditis [10], rheumatic heart disease [14] and, less frequently, Kawasaki disease [12], blunt chest trauma [7], acute rheumatic fever [2, 6], connective tissue disease [17, 22], and LV volume overload causing chorda stretching [14]. Although some reports have indicated that myxomatous valve disease is also a cause of primary chordal rupture [10, 16], this etiology remains speculative. It is unclear whether myxomatous degeneration is a primary cause or the result of mitral valve insufficiency.

Recent case reports on acute MR caused by chordal rupture in neonates describe an association with transplacental passage of the anti-SSA antibody [8, 21]. However, our cases were normally developed 4-month-old infants with acute MR caused by ruptured chordae tendineae showing no association with anti-SSA antibody. There are few case reports on ruptured chordae tendineae during childhood, and no infant series has been reported. We reviewed the clinical characteristics of ruptured chordae tendineae cases.

Methods

We reviewed the medical records of all patients diagnosed with acute MR due to ruptured chordae tendineae at the National Cerebral and Cardiovascular Center between December 2001 and June 2009. Diagnoses were MR and ruptured chordae tendineae, detected by transthoracic echocardiography (TTE) and later confirmed by intraoperative findings. Patients with congenital cardiac anomalies and Kawasaki disease were excluded.

We analyzed age at onset, sex distribution, symptoms at the first visit and upon admission to our institute, period of exacerbation, chest radiography, electrocardiograms (ECG), laboratory data, bacterial tests (preoperative blood culture, pharyngeal culture, and excised tissue culture), TTE findings (grade of MR, location of affected chordae, LV size and function, pulmonary hypertension, and vegetations), pathological findings, surgical procedures, and complications.

Hypertrophy of the atria and ventricles was defined using ECG; left atrial hypertrophy exhibited a broad (more than 0.10 s) bimodal P wave in leads I, II, and aVL, and a biphasic P wave with a negative prolonged latter portion in lead V1. LV hypertrophy exhibited a strain pattern of ST–T in leads V5 and V6, a high R wave (more than 2.5 or 3.5 mV in lead V6 or V5, respectively), and high total amplitude (more than 4.0 or 5.0 mV) of the S wave in lead V1 and the R wave in lead V6, or the S wave in lead V1 and the R wave in lead V5.

MR grading by TTE was classified based on extension of regurgitation on color Doppler imaging. Severe MR was defined as extension to pulmonary veins.

Statistical analysis

Continuous data are expressed as means±SD. Statistical analyses were performed using StatView for Windows version 5.0 (SAS Institute Inc, Cary, NC, USA). Comparisons between groups were conducted using the Fisher exact test. Differences were considered to be statistically significant at p values less than 0.05.

Results

Patient characteristics

Our study included ten consecutive patients (five boys and five girls) with acute MR due to ruptured chordae tendineae. Two patients with chordal rupture due to Bland-White-Garland syndrome and Kawasaki disease, respectively, were excluded. Mean age at onset was 4.6± 1.3 months. Most patients had no significant medical history, the exceptions being two patients with low birth weights and breath-holding spells (Table 1). There were no physical signs of connective tissue diseases (e.g., Marfan or Ehlers–Danlos syndrome). All patients showed normal development and had no heart murmurs during routine medical examination in infancy or at an outpatient clinic. All had an uneventful familial history. All patients had been transferred to our institute for further intensive treatment due to congestive heart failure with severe MR, as detected by TTE.

Symptoms at the first visit and on admission to our institute

During the first visit to previous hospitals, most patients showed nonspecific symptoms, including common cold-like symptoms, such as cough, fever, rhinorrhea, and poor feeding (Fig. 1). Only three exhibited pallor during the initial visit. These three patients were immediately administered inotropic agents and mechanical ventilation within 24 h of the first visit.

The mean duration between the first visit and admission to our hospital was 6.2±5.4 days. When transferred to our hospital, all patients had respiratory distress (Fig. 1), six were on mechanical ventilation, and five had received inotropic agents for circulatory insufficiency. Cardiopulmonary resuscitation (CPR) was provided for three patients before transfer. One patient required resuscitation upon arrival at our hospital. Despite these aggressive medical interventions, all patients developed respiratory distress within a few days of onset (mean, 1.8±1.8 days), which then progressed to circulatory insufficiency.

Table 1 Patients diagnosed as having acute mitral regurgitation due to chordal rupture

Age (month)	Mean, 4.6±1.3 (range, 2.0–6.6)
Sex (boy/girl)	5:5
Weight (g)	Mean, 6,439.2±1,349.6 (range, 3,944–8,320)
History	None, 8
	Low birth weight, 2
	Breath-holding spells, 1

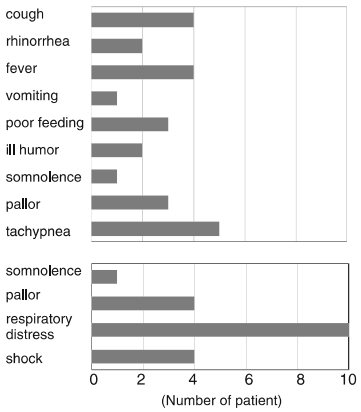

Fig. 1 Symptoms of acute mitral regurgitation due to chordal rupture at the first visit (*upper figure*) and on admission to our institute (*lower figure*). Note that six patients were already mechanically ventilated under sedation at the time of admission

Chest radiography findings

Chest radiographs showed pulmonary congestion in all ten patients. The cardiothoracic ratio was normal or mildly increased. Seven patients had cardiothoracic ratios under 60% (Table 2).

ECG findings

One patient had tall P waves in leads II, III, and aVF, but not in lead V1. None of the other patients had findings of atrial or ventricular hypertrophy, prolonged PR interval, arrhythmias, or ST–T changes (Table 2).

Laboratory data

Table 3 shows the laboratory data of all ten patients. Five had leukocytosis (>15,000/μL), while the others had white blood cell counts within normal range. C-reactive protein was within normal range (<0.5 mg/dL) in four patients and elevated in six. Creatine phosphokinase was slightly elevated in three patients. However, two of these patients had been administered with CPR before blood tests, and none of the three had evidence of ischemia on ECG or TTE. Human atrial natriuretic peptide (hANP) and brain natriuretic peptide (BNP) were markedly elevated in all patients. Three patients tested negative for anti-streptolysin O. Pharyngeal cultures were negative for group A *Streptococcus* in all patients.

Autoantibodies, including antinuclear, anti-ds DNA, anti-ss DNA, anti-RNP, anti-sm, anti-SS-A, and anti-SS-B antibodies, were measured. Only one patient had slightly

Table 2 Chest radiography, echocardiography, operative procedures, resuscitation, and complications

Patient	Sex	Onset (month)	CTR (%)	Pulmonary congestion	ECG	MR grade	Location of chorda	LVDd (mm, %)	LVEF	Operative procedure	CPR	Complications
1	F	2.0	65	+	–	Severe	Bilateral	22.3 (114.0)	0.79	Repair	+	CNS
2	M	6.6	47	+	Tall P	Severe	Anterior	30.0 (115.6)	0.94	Repair	+	CNS
3	F	5.3	67	+	–	Severe	Bilateral	28.5 (121.0)	0.83	Repair	–	–
4	M	4.9	59	+	–	Severe	Anterior→bilateral	27.0 (114.0)	0.87	Replace	–	–
5	M	3.7	60	+	–	Severe	Bilateral	28.1 (114.0)	0.86	Replace	–	AT
6	F	3.7	57	+	–	Severe	Anterior	21.4 (96.0)	0.81	Replace	+	AT, CNS
7	M	5.5	68	+	–	Severe	Posterior	27.2 (112.0)	0.76	Repair	–	–
8	F	6.2	60	+	–	Severe	Anterior	29.1 (131.0)	0.82	Repair→replace	–	SSS
9	F	4.2	53	+	–	Severe	Posterior	27.6 (116.0)	0.76	Repair	+	–
10	M	4.7	57	+	–	Severe	Anterior	30.2 (122.0)	0.74	Replace	–	AFL

LVDd left ventricular diastolic dimension; *LVEF* left ventricular ejection fraction; *CPR* cardiopulmonary resuscitation before surgery; *Tall P* tall P wave in leads II, III, aVF; *Repair* mitral valve repair; *Replacement* mitral valve replacement; *CNS* central nervous system damage; *AT* atrial tachycardia; *SSS* sick sinus syndrome; *AFL* atrial flutter

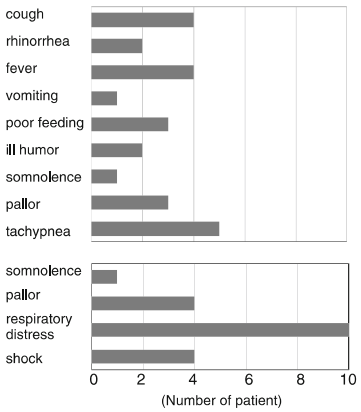

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Table 3 Average laboratory values on admission

WBC (per μL)	18,530±8,620 (range, 7,600–33,000)
Hb (g/dL)	8.7±2.2 (range, 6.5–14.4)
Ht (%)	25.8±6.1 (range, 19.5–41.9)
Plt (per μL)	46.1±17.1 (range, 25.7–70.1)
CRP (mg/dL)	2.0±2.5 (range, 0.1–7.8)
AST (IU/L)	274±634 (range, 22–2,067)
ALT (IU/L)	153±224 (range, 7–693)
LDH (IU/L)	1,026±1,373 (range, 181–4,584)
BUN (mg/dL)	11.2±5.7 (range, 5–20)
Cre (mg/dL)	0.35±0.17 (range, 0.1–0.7)
CK (IU/L)	1,045±2,714 (range, 27–8,755)
ANP (pg/mL)	2,377±2,819 (range, 376–7,290)
BNP (pg/mL)	2,322±2,158 (range, 341–6,026)

WBC white blood cell, *Hb* hemoglobin, *Ht* hematocrit, *Plt* platelet, *CRP* C-reactive protein, *AST* aspartate aminotransferase, *ALT* alanine aminotransferase, *LDH* lactate dehydrogenase, *BUN* blood urea nitrogen, *Cre* creatinine, *CK* creatine phosphokinase, *ANP* atrial natriuretic peptide, *BNP* brain natriuretic peptide

elevated anti-ds DNA and anti-ss DNA antibodies; in all other patients, these autoantibodies were undetectable.

Bacterial culture

Preoperative blood cultures were obtained in eight patients, half of these before antibiotic administration. All specimens were negative. Pharyngeal cultures showed normal flora.

Echocardiography

TTE showed severe MR with ruptured chorda in all patients (Table 2; Fig. 2). Affected mitral valve leaflets fluttered widely and protruded into the left atrium during systole. These features differed from those of mitral valve prolapse cases. The location of affected chordae tendineae was anterior in five patients, posterior in two, and both in three. Interestingly, one patient with anterior chordal rupture showed sudden deterioration of respiratory distress on the second day of hospitalization. TTE showed progression of MR due to further rupture of the posterior chordae tendineae.

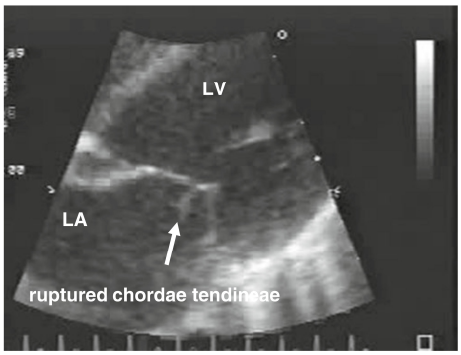

Fig. 2 Preoperative transthoracic echocardiogram from patient 9. This patient had respiratory distress and perfusion failure at the first visit and had been intubated at the previous hospital; the patient was transferred to our institution the same day. Posterior ruptured chorda was detected (*arrow*)

In all patients, the LV diastolic dimension (LVDd) was normal or mildly enlarged. The mean LVDd was 27.1±2.9 mm, i.e., 115.6±8.8% of normal values as calculated from body surface area. LV ejection fractions were normal (>0.65), and no ventricular asynergy was detected. Five patients had mild pulmonary hypertension, as estimated by tricuspid valve regurgitation flow velocity.

Operation

All patients required surgical intervention for mitral valve repair. Eight of the ten (80%) underwent emergency surgery within 24 h of admission, and the mean period from admission to the operation was 3.6±5.1 h (range, 0.6–16.0 h). Two patients underwent surgery the day after admission. One patient with further posterior chordal rupture on the second day of hospitalization required emergency surgery. Another patient experienced brain hemorrhage, a contraindication for open-heart surgery. He underwent mitral valve repair after stabilization following the acute phase of hemorrhage.

Mitral valve repair, including Kay annuloplasty, edge-to-edge repair, and artificial chorda, was conducted in six patients. One subsequently required mitral valve replacement due to further chordal rupture postoperatively. Five patients underwent mitral valve replacement using 16-mm ATS AP series mechanical valves (ATS Medical Inc., MN, USA) translocated to the supraannular position (Table 2).

Intraoperative findings revealed that all patients had experienced chordal rupture and four had yellow or yellowish-white degeneration at the ruptured chordae; in three of these four, this degeneration extended to the mitral leaflet edge. There were no other abnormalities, such as vegetations, ischemia, infection, and commissural fusion.

In all patients, surgical treatment improved cardiopulmonary insufficiency. Inotropic agents and mechanical ventilation were discontinued. Postoperative echocardiography showed amelioration of MR to mild or moderate in the mitral valve repair group and no cases with mitral valve replacement experienced leakage. The hANP and BNP levels decreased markedly to 164±27.4 and 90±66.7 pg/mL, respectively.

Pathological findings

Pathological examination of excised tissues revealed six specimens to have infiltration of inflammatory cells, including mild lymphocyte or neutrophil and macrophage infiltration, while four showed fibrin adhesion. Myxomatous degeneration was noted in two specimens, fibrosis in one. There was no evidence of bacterial infection in any of the specimens. We stained for acid-fast bacteria to investigate the possible relationship to Bacillus Calmette–Guérin vaccination in two specimens, but the results were negative.

Complications

Three of the four patients who required CPR exhibited complex central nervous system damage. The first patient had a convulsion after the operation and was successfully managed using anticonvulsants. As of the latest follow-up, this patient exhibits no developmental delays. The second patient had brain hemorrhage, brain atrophy, and developmental retardation. The third patient had hypoxic encephalopathy, subdural hematoma, and developmental retardation.

Four patients developed atrial arrhythmias after mitral valve replacement. The mitral valve replacement group had a much higher rate of atrial arrhythmias (80%) than the mitral valve repair group (0%; $p<0.05$). Atrial tachyarrhythmias occurring in three patients were all well controlled using anti-arrhythmic agents. One patient required pacemaker implantation due to sinus node dysfunction. In this patient, postoperative ECG showed elevated ST segments in leads V5, V6, II, III, and aVF. TTE showed hypokinesis at the LV inferior wall. We suspected that the prosthetic valve compressed the left circumflex artery (LCX), causing hypoperfusion of the sinus node artery arising from the LCX. Aortography performed after surgery confirmed that the sinus node artery arose from the LCX (Fig. 3).

Discussion

In the present study, we reviewed the clinical characteristics of idiopathic acute MR due to ruptured chordae tendineae in infancy. Symptom onsets were mostly in early infancy, at approximately 4 months of age, with no significant difference in sex distribution. Initial symptoms were nonspecific, resembling those of a common cold. However, all patients developed respiratory distress within a few days and most progressed to circulatory failure despite aggressive medical intervention. Chest radiographs revealed pulmonary congestion, even though the cardiothoracic ratio was normal or only mildly increased. ECGs revealed no atrial hypertrophy, ventricular hypertrophy, or ST–T changes. In some patients, laboratory data indicated inflammation, but this was usually not significant. These nonspecific findings could easily be misinterpreted as representing lung disease. Indeed, some cases in our series were initially misdiagnosed and treated for pneumonia or acute respiratory distress syndrome. Therefore, we advocate performing echocardiography when an infant presents with a vague set of symptoms, i.e., "not doing well," and shows pulmonary congestion on chest radiography.

The major causes of acute MR due to chordal rupture in adult series are reportedly ischemia [13, 14], infectious endocarditis [10], and rheumatic heart disease [11, 14]. However, these etiologies are rare in childhood. Previous reports on acute MR during childhood are limited and include causes such as Kawasaki disease [12], acute rheumatic fever [2, 6], autoimmune disorders [8, 21], connective tissue diseases [22], and chest trauma [3]. However, these cases had known causative factors. We carefully ruled out all known causes of acute MR in our series. Therefore, the cases presented herein appear to be idiopathic.

There are few reports on the pathological findings of ruptured chordae tendineae during childhood. In our series, some patients showed inflammatory cell infiltration, including lymphocyte or neutrophil and macrophage

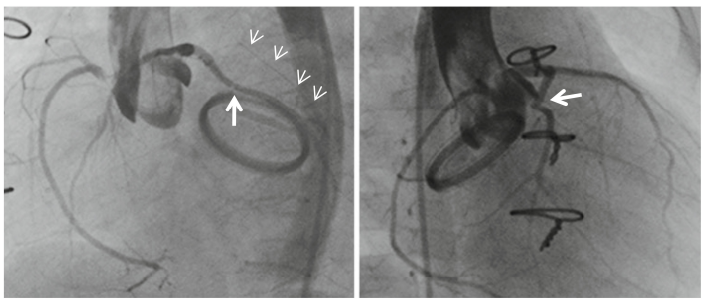

Fig. 3 Aortography after mitral valve replacement in patient 9. Sick sinus syndrome and ischemic change on electrocardiography and echocardiography were recognized after surgery. The prosthetic valve compressed the left circumflex artery from which the sinus node artery (*small arrows*) originated (*arrow*)

infiltration, or myxomatous degeneration, on pathological specimens. Inflammatory cell infiltration is generally caused by infection, autoimmunity, or other immunological mechanisms, including mechanical inflammation. However, there was no evidence of bacterial infection based on either cultures or pathology results. Moreover, levels of infant serum autoantibodies that could damage chordae tendineae, such as maternal-derived anti-SSA antibody [8], were low or undetectable. A few case reports on chordal rupture associated with anti-SSA antibody described fibrotic scarring and calcification as characteristic intraoperative findings, but histological studies were not conducted. The literature on chordal rupture in adult cases describes myxomatous degeneration as one of the causes of primary chordal rupture [10, 16]. The mechanical strength of the mitral leaflet and chordae tendineae was shown to be decreased because of the matrix remodeling, due in part to both dysregulation of matrix components and abnormal mechanical stress, but the primary stimulus for this degeneration was unclear [9, 15]. Degeneration is primarily idiopathic and may result from age-related changes or excessive hemodynamic loads for prolonged periods. Therefore, it is not reasonable to assume that these effects cause chordal rupture in early infancy. In our series, myxomatous degeneration was a nonspecific finding.

Detailed examinations revealed no evidence of bacterial infection, autoimmune disease, acute rheumatic fever, Kawasaki disease, connective tissue disease, congenital heart disease, or chest trauma in any of the ten cases. Moreover, the onsets were in early infancy, generally at approximately 4 months of age, and the majority of patients initially had symptoms similar to those of the common cold. Hence, we suspect that age-dependent factors, including specific anatomical weaknesses of mitral valve structure and immunological responses associated with viral infections, may have contributed to the onset. Based on our present findings, acute MR due to chordal rupture in infancy is a definite disease entity that differs from chordal rupture in adults. This study was retrospective and did not reveal the origin of chordal rupture. Further prospective studies focusing on immunological mechanisms, such as infant responses to maternal-derived autoantibody and exogenous antigens including those associated with viral infections, are needed to clarify the etiology of this pediatric disease entity.

The anatomical mechanism underlying mitral chordal rupture is also largely unknown. However, a few studies have provided possible insights. Oliveira et al. reported that the major area of the anterior leaflet is tangential to systolic flow, as opposed to the posterior leaflet, which is perpendicular to flow [14]. Sedransk et al. described the marginal and posterior leaflet chordae as being thinner and requiring less strain and load to fail than the basal and anterior leaflet chordae, respectively [18]. Other studies involving adults have also noted that in patients with primary and secondary rupture, posterior rupture is most common [10, 14]. Our results regarding the locations of affected chordae tendineae are not consistent with these studies. This difference might be attributable to specific etiological factors, such as anatomical weaknesses, in infancy that differ from those in adults.

Although successful management with medical interventions only has been reported, most studies found surgery to be required in cases of acute MR during childhood [1, 3, 6, 8, 11, 21]. Early surgical intervention before the development of organ damage is important, particularly for preventing neurological sequelae. In this study, most cases underwent surgery within 16 h after emergency admission. Mitral valve repair is desirable to avoid the complications of prosthetic valve replacement. However, with leaflet edge involvement, the mitral valve is so fragile that repair may not be possible due to leaflet vulnerability. Mitral valve replacement in pediatric patients carries high risks of damaging the conduction system, compressing the LCX, obstructing the LV outflow tract, bleeding, thromboembolism, and general structural or non-structural prosthetic valve complications [4, 19, 20]. Moreover, particularly in infants, prosthetic valves must be placed in the supra-annular position in many cases because of the mismatch between prosthetic valve size and infant annulus size. Supra-annular placement is accompanied by a risk of pulmonary hypertension resulting from reduced left atrial volume and LV failure due to paradoxical motion of the atrium below the prosthetic valve [4, 19]. Fortunately, in our series, these complications had not occurred as of the most recent follow-up.

Study limitations

This was a retrospective study. A prospective study investigating etiologies, including immunological mechanisms and the causes of ruptured chordae tendineae in infants, is required.

Conclusions

Acute MR due to ruptured chordae tendineae in infancy is characterized by acute onset, rapid development of respiratory distress, and left heart failure, despite intensive medical treatment. Pulmonary congestion is a specific finding on chest radiographs even without significant cardiomegaly. Therefore, for early diagnosis, TTE is necessary when an infant appearing unwell shows pulmonary congestion. Early surgical intervention is recommended for good prognosis. Our data indicate no evidence of bacterial

infection or infant autoantibodies. The onsets were in healthy and well-developed infants, approximately 4 months of age. Thus, these factors suggest a definite disease entity, which should be recognized by not just pediatric cardiologists but also pediatric emergency physicians.

Conflicts of interest The authors have no financial interests in any of the treatments or devices described in this article. None of the authors has any conflicts of interest to declare.

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