Table 2. Risk and Precautions for Blood Component Transfusion in IgA-Deficient Patients

<table>
<thead>
<tr>
<th>Patients at risk for anaphylactic reactions related to IgA</th>
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<tr>
<td>IgA deficient (&lt;0.05 mg/dL)</td>
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<tr>
<td>High titers of anti-IgA antibody</td>
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<table>
<thead>
<tr>
<th>Precautions for patients at risk for anaphylactic transfusion reactions related to IgA</th>
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<tr>
<td>Washed blood products</td>
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<tr>
<td>Blood products from IgA-deficient donor</td>
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<tr>
<td>Recombinant clotting factor concentrates</td>
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IgA = immunoglobulin A.

viduals [7]. Since this frequency far exceeds the number of observed anaphylactic transfusion reactions, it is clear that detection of an anti-IgA antibody does not reliably predict risk of anaphylaxis [8]. Nevertheless, among patients who experience an anaphylactic transfusion reaction, documentation of IgA deficiency and an anti-IgA antibody may inform subsequent transfusion support, as this case illustrates.

Subsequent transfusion support for patients with a history of anaphylaxis caused by IgA deficiency should minimize reexposure to IgA contained in blood products or derivatives (such as plasma-derived clotting factors). The 2 principle methods used for blood products include washing blood components to remove IgA and obtaining blood components from donors known to be deficient in IgA (Table 2). Although red blood cells are easily washed to remove IgA, platelets are lost during the washing process and have reduced survival in the patient after transfusion. Plasma products such as FFP cannot be washed. Therefore obtaining these components from IgA-deficient donors, as done in this case, is preferred.

For patients with a history of a single episode of anaphylaxis associated with blood transfusion and subsequently found to have normal IgA levels, it may be reasonable to retransfuse with careful monitoring before restricting them to washed blood components.

Hemophilia A is a sex-linked recessive condition resulting in various levels of factor VIII [9]. Patients are at risk for significant bleeding during surgical procedures. Several different protocols, including intermittent bolus administration of recombinant factor VIII and/or continuous infusion of factor VIII, are commonly used. In general, factor VIII administration is continued well into the postoperative period. Rossi and colleagues [9] performed a review of reported cardiac surgical cases in patients with hemophilia and suggested that good surgical outcomes are possible with preoperative planning and a team approach to management. Recombinant factor VIII preparations have no IgA and are thus safe for transfusion to patients with high-titer anti-IgA antibodies.

This is the first report of successful management of a complicated thoracic surgical procedure requiring repeated sternotomy in a patient with both a history of hemophilia A and an anaphylactic transfusion reaction related to IgA deficiency. Management of such complicated cases is successful when previous planning and a team approach is undertaken.

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References


Falsely Elevated Valve Gradients by Echocardiography in the 3f Aortic Bioprosthesis

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The 3f Aortic Bioprosthesis (Medtronic, Inc, Minneapolis, MN) is a stentless aortic valve with a novel design that resembles a “tube within a tube.” Although it has the potential for improved durability and hemodynamic performance, long-term data on this valve remain elusive. We present here 3 patients in whom postoperative echocardiography revealed significantly elevated transvalvular gradients of the 3f valve while transcatheter gradients proved to be negligible. By virtue of the unique design of the 3f bioprosthesis, great caution should be taken when interpreting echocardiographically derived gradients.


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The 3f Aortic Bioprosthesis (Medtronic, Inc, Minneapolis, MN) is a stentless equine pericardial valve that has been in use for the past 4 years in North America, with earlier experience in Europe. The 3f valve design allows simplified implantation and has garnered good early hemodynamic results in small single-center studies [1–3]. However, recent midterm data have shown less favorable echocardiographic hemodynamic results in the smaller prosthesis sizes [4]. Although transthoracic echocardiography (TTE) is a useful tool in assessing prosthetic valve function, it can present extraneous results. We present here 3 cases in which falsely elevated transvalvular gradients were demonstrated by echocardiography in the 3f aortic valve.

Case Reports

Case 1
A 79-year-old woman underwent aortic valve replacement (AVR) with a 19-mm aortic bioprosthesis in November of 2010 for severe aortic stenosis and New York Heart Association (NYHA) class II symptoms. Preoperative TTE demonstrated an aortic valve area of 0.7 cm² and an aortic valve mean systolic gradient of 34 mm Hg, with a mildly decreased left ventricular ejection fraction (LVEF) of 45% to 50%. Coronary angiography was normal and her body surface area was 1.40 m².

Postoperatively, the patient recovered well and was discharged home. The patient returned to the hospital two months later with symptoms of orthopnea, peripheral edema, and atrial fibrillation. Repeated TTE showed a peak velocity of 3.8 m/sec, a peak/mean aortic systolic pressure gradient of 40 mm Hg, an aortic valve area of 1.05 cm², and a normal LVEF of 50%. Although transthoracic echocardiography (TTE) is a useful tool in assessing prosthetic valve function, it can present extraneous results. We present here 3 cases in which falsely elevated transvalvular gradients were demonstrated by echocardiography in the 3f aortic valve.

Case 2
A 53-year-old woman underwent AVR with a 21-mm 3f aortic bioprosthesis in May 2008 for NYHA class II heart failure symptoms. Preoperative TTE demonstrated a morphologically normal aortic bioprosthesis and normal biventricular function but a peak velocity of 3.64 m/sec, a peak/mean aortic systolic pressure gradient of 53/27 mm Hg, a DVI of 0.33, and an EOA of 1.12 cm² (0.59 cm²/m²) by the continuity equation, suggestive of possible stenosis. However the contour of the jet velocity appeared triangular with an early peak. History taking revealed that the patient was asymptomatic. These elevated gradients were postulated to be related to high transvalvular flow rates (cardiac index 3.4 L/min/m²).

At 6-month follow-up, repeated TTE was virtually unchanged from the previous, with a peak velocity of 3.17 m/sec, a peak/mean aortic valve gradient of 50/25 mm Hg, and an EOA of 1.14 cm² (0.60 cm²/m²), with a cardiac index of 3.5 L/min/m². Continuous wave Doppler examination of the aortic valve displayed indentation artifacts on the contour and through the Doppler signal (Fig 3). In addition, at this time the patient experienced peripheral edema of the upper and lower extremities.

Dr de Varennes discloses a financial relationship with Medtronic.
and NYHA class II symptoms. In the context of recently discontinuing hormone treatment for menopause, it was difficult to decipher an endocrine versus cardiac cause for these symptoms. Given consistent echocardiographic evidence of aortic stenosis, invasive evaluation of the prosthetic valve was performed. At 18 months postoperatively, she underwent a cardiac catheterization that demonstrated normal LVEF and no significant gradient across the prosthetic valve by the pullback method (Fig 4). The patient subsequently resumed her hormone therapy, and her symptoms resolved on follow-up.

**Case 3**

A 37-year-old man underwent aortic root replacement for a dilated aortic root and bicuspid aortic valve. On history taking, the patient had NYHA class II dyspnea and Canadian Cardiovascular Society class II angina. Initial workup demonstrated a dilated ascending aorta measuring 5.6 cm, a dilated aortic root of 4.1 cm, moderate aortic regurgitation from a bicuspid aortic valve but preserved LVEF on TEE. A coronary angiogram revealed normal coronary anatomy and no atherosclerosis.

The patient underwent a modified Bentall procedure with a 29-mm 3f Aortic Bioprosthesis implanted within a 30-mm Vascutek Gelweave Valsalva graft (Terumo, Ann Arbor, MI) in May 2010. Body surface area at the time of operation was 2.08 m². The patient's postoperative course was uneventful and he was discharged home on postoperative day 5.

Three-month postoperative TTE showed a peak aortic valve velocity of 3.12 m/sec, peak/mean transvalvular gradients of 39/20 mm Hg, and an EOA of 2.2 cm² (1.04 cm²/m²) with normal LVEF. Again, the contour of the maximal jet velocity appeared triangular with early peaking. TTE 1-year postoperatively produced the same elevated transvalvular gradients but normal left ventricular mass. On echocardiographic review, continuous wave Doppler showed high-frequency indentation artifacts associated with demonstrated high-speed fluttering of the aortic leaflets on M-mode recordings at high sweep speed (Fig 5). Given these findings, no further invasive testing was performed with regard to the hemodynamic profile of the bioprosthesis.

**Comment**

Doppler and TTE examination are the modalities of choice for noninvasive evaluation of prosthetic valve function because they provide accurate hemodynamic data [5]. We have implanted more than 200 3f valves to date and reveal here the difficulties of assessing this prosthetic valve by echocardiography in some patients. Given the novel design of the 3f aortic valve, Cox and associates [6] demonstrated improved flow dynamics and prosthesis stress distribution in an in vitro model. However, we have shown in this case series that in vivo the
The design of this tissue valve may cause erroneously elevated Doppler gradients by high-frequency fluttering of the replacement leaflets. As shown in Figs 3 and 5 in 2 distinct patients, this fluttering seen on M-mode with corresponding artifacts on the Doppler signal raises the question of whether tracing the outside contour of the Doppler signal reflects the true red blood cell maximal velocity used for the measurement of DVI and estimation of EOA by the continuity equation.

Other pitfalls leading to erroneous aortic valve gradients by Doppler include misalignment of the ultrasound beam with the flow through the aortic valve, as well as mistaking other high-velocity jets for the aortic jet [7]. In addition, the phenomenon of pressure recovery has also been used to explain the overestimation of Doppler gradients compared with gradients obtained by catheterization. Pressure recovery occurs when the velocity of the ejected blood decreases and this kinetic energy is converted to potential energy, which is seen as a rise or recovery in the static pressure. The pressure gradient between the left ventricle and the aorta after pressure recovery is the pressure gradient of interest when interpreting the hemodynamic performance of a prosthetic valve and that measured by catheterization. Several studies have demonstrated pressure recovery in stented aortic prosthetic valves, although not yet in stentless valves [8].

The lack of long-term data on this valve poses a great challenge in assessing its hemodynamic performance. We are currently in the process of gathering echocardiographic data on our entire cohort of 3f valves, which should help determine whether the observations in this series are the exception or the norm. The results of the 3 cases presented here should, however, provide caution when obtaining a Doppler-derived gradient that is greater than expected. If the clinical situation warrants it, transcatheter measurements of the true gradients should be obtained before any decision that might affect the surgical management of patients with the 3f valve.

References

Fig 4. Case 2. Left heart catheterization. Peak-to-peak aortic systolic gradient on pullback into the ascending aorta is near 0 mm Hg.

Fig 5. Case 3. (A) M-mode recording of the 3f bioprosthesis from parasternal long-view axis window by transthoracic echocardiography (TTE). Note the high-frequency fluttering of the prosthetic aortic valve leaflets (red arrow). (B) Continuous wave Doppler interrogation of the bioprosthetic valve from the apical long-axis view demonstrating concomitant high-frequency indentations (red arrow) through the Doppler signal.
Severe pulmonary hypertension is associated with a poor prognosis [1]. Chronic suprasystemic pulmonary hypertension and right ventricular pressure overload usually result in right ventricular failure [2]. A decrease of cardiac output is not only related to impaired right ventricular function but also could be a consequence of impaired left ventricular filling due to right-to-left ventricular interaction [3] or reduced left ventricular function, ie, due to a borderline left ventricular morphologic condition [4]. As a novel approach for palliating children with suprasystemic idiopathic pulmonary arterial hypertension (IPAH), creation of a Potts shunt has been reported, which could be an alternative treatment to lung transplantation in selected cases [4, 5]. This surgical procedure implies the construction of an anastomosis between the left pulmonary artery and the descending aorta that allows right-to-left shunting and thereby leads to decompression of the failing right ventricle (RV) without provoking desaturation in the upper part of the body.

We describe our experience with a modified Potts shunt in a 20-year-old young adult with suprasystemic postcapillary pulmonary hypertension caused by Shone’s complex, including parachute mitral valve, aortic valve stenosis, small muscle ventricular septal defect, persistent ductus arteriosus, and a LV of borderline size. The patient had successful balloon valvuloplasty of the stenotic aortic valve at the age of 1 month and an interven- tional closure of the arterial duct at the age of 2 years. At the age of 20 years he was admitted to our center with decompensated biventricular failure corresponding to World Health Organization functional class IV and was already listed for combined heart-lung transplantation. Horizontal positioning became impossible and resulted in acute congestive heart failure associated with pulmonary edema and significant cyanosis despite continuous supplemental oxygen therapy and pulmonary arterial hypertension (PAH)-specific medication that included a combination of bosentan and sildenafil.

Cardiac imaging by echocardiography/cardiac magnetic resonance imaging showed a small LV with impaired function (indexed end-diastolic left ventricular volume, 77 mL/m² body surface area; mitral valve annulus, 31 mm; ejection fraction, 34%), an enlarged left atrium, and dilated pulmonary veins. The apex-forming RV and the pulmonary arteries were markedly dilated, the right ventricular myocardium was hypertrophied, and right ventricular function was significantly impaired (ejection fraction, 16%) (Figs 1, 2). Right and left heart catheterization performed in the sitting position revealed low cardiac output (cardiac index, 2.2 L/min/m²; Qp/Qs [ratio of pulmonary blood flow to systemic blood flow], 1.0) associated with severe left ventricular diastolic dysfunction (left ventricular end-diastolic pressure, 28 mm Hg) and “suprasystemic” postcapillary pulmonary hypertension (pulmonary arterial pressure, 125/75 mm Hg; systemic arterial pressure 105/55 mm Hg). Nitric oxide administration...