JACC: CASE REPORTS VOL. 2, NO. 1, 2020


PERIPARTUM CARDIOVASCULAR DISEASE MINI-FOCUS ISSUE

ADVANCED

CASE REPORT: CLINICAL CASE

# Two Hearts at Risk

## Emergency Alcohol Septal Ablation in a Pregnant Woman With Decompensated HOCM

Weng-Tein Gi, MD, MSc,[a,b] Ali Amr, MD,[a,b] Farbod Sedaghat-Hamedani, MD,[a,b] Elham Kayvanpour, MD,[a,b] Isabell Mohr, BSc,[a] Manuela Meder, MD,[a] Omid Shirvani Samani, MD,[a,b] Herbert Fluhr, MD,[c] Hugo A. Katus, MD,[a,b] Benjamin Meder, MD[a,b,d]

**ABSTRACT**

Hypertrophic obstructive cardiomyopathy (HOCM) increases the risk for mother and fetus during pregnancy. Alcohol septal ablation (ASA) is an established procedure in nonpregnant patients with HOCM. In this report, we present a case of a 29-year-old woman in her 29th gestational week with decompensated HOCM undergoing a successful ASA. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2020;2:139–44) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

### HISTORY OF PRESENTATION

A 29-year-old pregnant patient in her 29th week of gestation was referred to the intermediate care unit of our gynecology university clinic due to shortness of breath. She reported severe dyspnea after minimal exertion (New York Heart Association functional class III) and was hypotensive with a blood pressure of 95/60 mm Hg. Her heart rate was 100 beats/min. She had no chest pain, abdominal pain, or signs of infection. Her physical examination revealed a 3/6 mid-systolic murmur over the lower left sternal border and fine crackles over both lower lungs. She also exhibited mild pretibial pitting edema.

**LEARNING OBJECTIVES**

- ASA can be safely performed in pregnant patients with HOCM to achieve rapid improvement of hemodynamics and functional class if conservative treatment fails.
- H(O)CM increases the risk for mother and child during pregnancy.
- During the prenatal consultation, proactive invasive gradient reduction strategies before planned pregnancies should be considered in high-risk patients with HOCM.

### MEDICAL HISTORY

The patient had a known, mostly asymptomatic hypertrophic obstructive cardiomyopathy (HOCM). She received an implantable cardiac defibrillator as primary prophylaxis 7 years ago and had previously given birth to 1 daughter without complications. The last echocardiogram before pregnancy showed a mild left ventricular outflow tract (LVOT) obstruction with an interventricular septum (IVS) thickness of 38 mm and a gradient of 36 mm Hg, both at rest and after Valsalva maneuver. During her early pregnancy, no echocardiograms were performed, nor was she admitted to a specialized center for observation. At 24 weeks of gestation, her yearly echocardiogram was performed, which demonstrated an increased LVOT

From the [a]Institute for Cardiomyopathy, Department of Medicine III, University of Heidelberg, Heidelberg, Germany; [b]DZHK (German Centre for Cardiovascular Research), Heidelberg/Mannheim, Germany; [c]Department of Obstetrics and Gynecology, Heidelberg University Hospital, Heidelberg, Germany; and the [d]Department of Genetics, Stanford University, Stanford, California.
All authors have reported that they have no relationships relevant to the contents of this paper to disclose.

Informed consent was obtained for this case.

Manuscript received October 15, 2019; revised manuscript received November 25, 2019, accepted November 25, 2019.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2019.11.053

**ABBREVIATIONS AND ACRONYMS**

**ASA** = alcohol septal ablation / transcoronary ablation of septal hypertrophy

**HCM** = hypertrophic cardiomyopathy

**HOCM** = hypertrophic obstructive cardiomyopathy

**IVS** = interventricular septum

**LV** = left ventricle

**LVOT** = left ventricular outflow tract

**LVOTO** = left ventricular outflow tract obstruction

**SAM** = systolic anterior motion

obstruction (LVOTO) with a gradient of 58 mm Hg at rest and a gradient of 108 mm Hg after Valsalva maneuver. Four weeks later, the patient developed pronounced symptoms and had a maximal LVOT gradient of 121 mm Hg. Hence, she was promptly referred to our hospital.

## DIFFERENTIAL DIAGNOSIS

The differential diagnosis included acute pulmonary embolism, acute pulmonary edema, or decompensated HOCM.

## INVESTIGATION

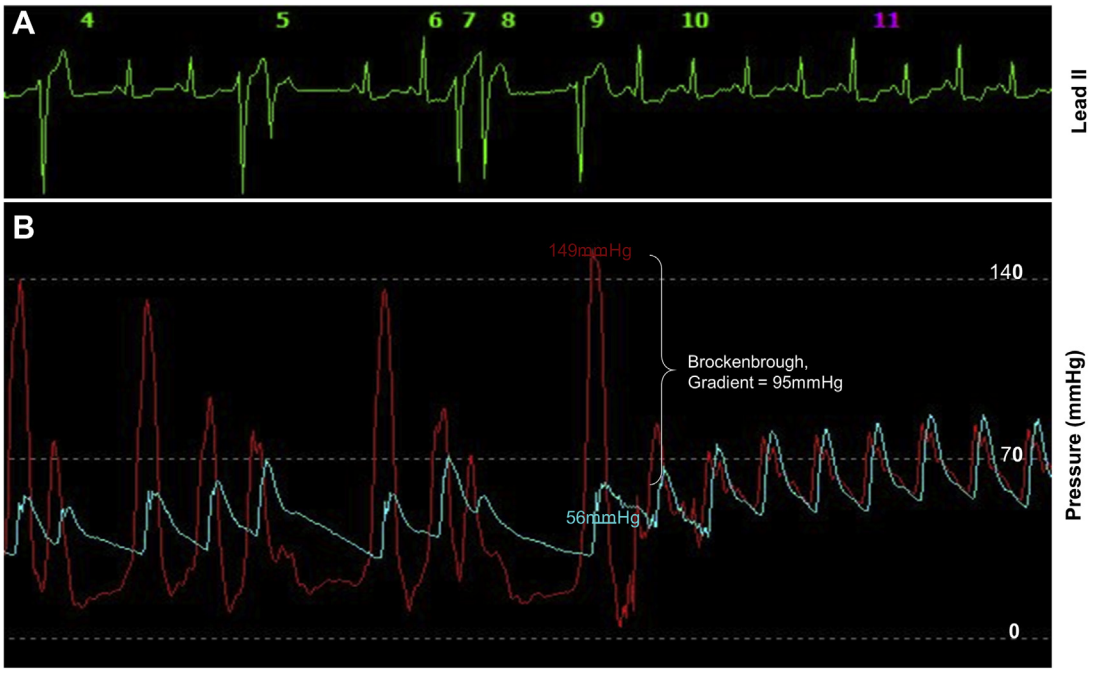
A 12-lead electrocardiogram revealed tall R waves in leads I and aVL as well as deep S waves in leads $V_2$ and $V_3$, suggesting hypertrophy of the left ventricle (LV). There were also T-wave inversions in leads II, III, aVF, and V6. Her echocardiogram showed an increased LVOT gradient of 65 mm Hg at rest, accompanied by an obvious systolic anterior motion of the mitral valve (SAM), which led to severe mitral regurgitation. The basal IVS was noticeably hypertrophied with a thickness of 38 mm. Her blood tests revealed markedly elevated N-terminal pro-B-type natriuretic peptide (3,965 ng/l) in the plasma. Based on these findings, the patient was diagnosed with acutely decompensated HOCM.

## MANAGEMENT

The patient received steroid therapy to accelerate fetal lung maturation. After our cardiomyopathy team was informed and the patient was seen at bedside, the therapy with Metoprolol succinate was uptitrated to 237.5 mg/day (95 mg [morning], 47.5 mg [noon], 95 mg [evening]). We carefully evaluated the use of diuretics and decided against it, because the patient was hypotensive and tachycardiac, indicating imminent cardiogenic shock (shock index >1.1) and she had a very narrow LV cavity with severe outflow tract obstruction. In such cases, diuretics and other afterload-changing drugs could result in increased gradients and hemodynamic instability. Instead of using diuretics, the patient's fluid intake and spontaneous diuresis were carefully balanced. Two echocardiographies were performed daily to monitor her LVOT gradient and vena cava inferior filling. Despite the ongoing therapy, her symptoms worsened rapidly. On the next day after admission, the patient developed orthopnea due to severe pulmonary edema, and required 8 liters of oxygen per minute to maintain an adequate oxygenation. Tachycardia and hypotension persisted. The edema in her lower extremities worsened. Her N-terminal pro-B-type natriuretic peptide surged to 8,748 ng/l. Furthermore, sonography showed a delayed intrauterine growth development of the fetus. Thus, an emergent cesarean delivery was considered by the gynecologists. To determine the ultimate management of the patient, the Institute for Cardiomyopathies Heidelberg organized an ad hoc interdisciplinary team discussion. After a comprehensive discussion among interventional cardiologists, gynecologists, anesthesiologists, and neonatologists, we decided to perform an urgent alcohol septal ablation (ASA). This was based on the fact that the conservative treatment was ineffective, our center has a high level of expertise in the ASA procedure with no cases of intrahospital death or anterior wall infarction, and an emergent cesarean delivery with (general) anesthesia could have resulted in hemodynamic instability. In addition, the patient was protected against heart block by her implantable cardiac defibrillator. The patient and her family were thoroughly informed about the ASA procedure and about the fact that to that date no published cases or recommendations concerning her medical situation existed. The family agreed to the procedure.

The ASA procedure was performed in a hybrid catheter laboratory, which was fully equipped in case of an emergency cesarean delivery. Aside from the cardiologists and catheter technicians, the treating gynecologist, an obstetrician, a neonatologist nurse, and an anesthesiologist were at the scene during the procedure. Not only the mother's but also the fetus's vital signs were continuously monitored. Before septal ablation, we hemodynamically measured the LVOT gradient by placing a pigtail catheter in the LV cavity (**Figure 1**) and performed a coronary angiogram to judge the anatomic suitability for ASA (**Figure 2A**). The total dosage of radiation during the complete procedure was only 0.75 mGy with the additional protection of the fetus by optimal radiation shielding. After selective occlusion of the first septal branch with an over-the-wire balloon (2-mm diameter, 6 psi occlusion pressure) (**Figure 2B**), we tested for retrograde leakage by applying contrast agent via the balloon's lumen (**Figure 2C**) and verified the targeted region by selective contrast echocardiography with 1 ml Sonovue contrast agent also given via the balloon lumen. As seen in **Figure 3B**, the contrasted region corresponded to the basal septum, which we aimed to ablate. Based on the contrasted target region, we planned to use 2.5 ml of 99% medical alcohol (0.1 ml/mm myocardium). Then, under continuous monitoring of cardiac electrical conduction and

**FIGURE 1** Electrocardiogram and Hemodynamic Monitoring During ASA




**(A)** Real-time electrocardiogram monitoring showing ventricular extra beats induced by the insertion of the pigtail catheter into the left ventricle. **(B)** Simultaneous monitoring of left ventricular pressure **(red line)** and aortic pressure **(blue line)** during catheterization. Induced premature ventricular contractions worsened the LVOT obstruction with a maximal gradient of 93 mm Hg (Brockenbrough-Braunwald-Morrow sign). ASA = alcohol septal ablation; LVOT = left ventricular outflow tract.

hemodynamics, we slowly injected the alcohol over 5 min via the lumen of the balloon. The patient received opioids at a tolerable dose to treat her angina pectoris and to slightly sedate the fetus. The fetus was constantly monitored by cardiotocography throughout the whole procedure, showing no signs of accelerations or decelerations and the peri-interventional sonographic examination of the fetus showed unremarkable findings.

One hour after the procedure, the patient's dyspnea improved considerably and the oxygen insufflation was reduced to 2 l/min. In her echocardiogram on the next day, the LVOT gradient had decreased from 65 mm Hg to 16 mm Hg, and only minor SAM of the mitral valve and trace mitral regurgitation could be seen (**Figure 4**). Pericardial effusion or a ventricular septum perforation were excluded. The patient's 12-lead electrocardiogram presented a new complete right bundle branch block, but no atrioventricular blockage. The patient was mobilized the next day, and was transferred to the normal ward 2 days later without any limiting dyspnea (New York Heart Association functional class I). Six days after the ASA procedure, the patient was discharged home.

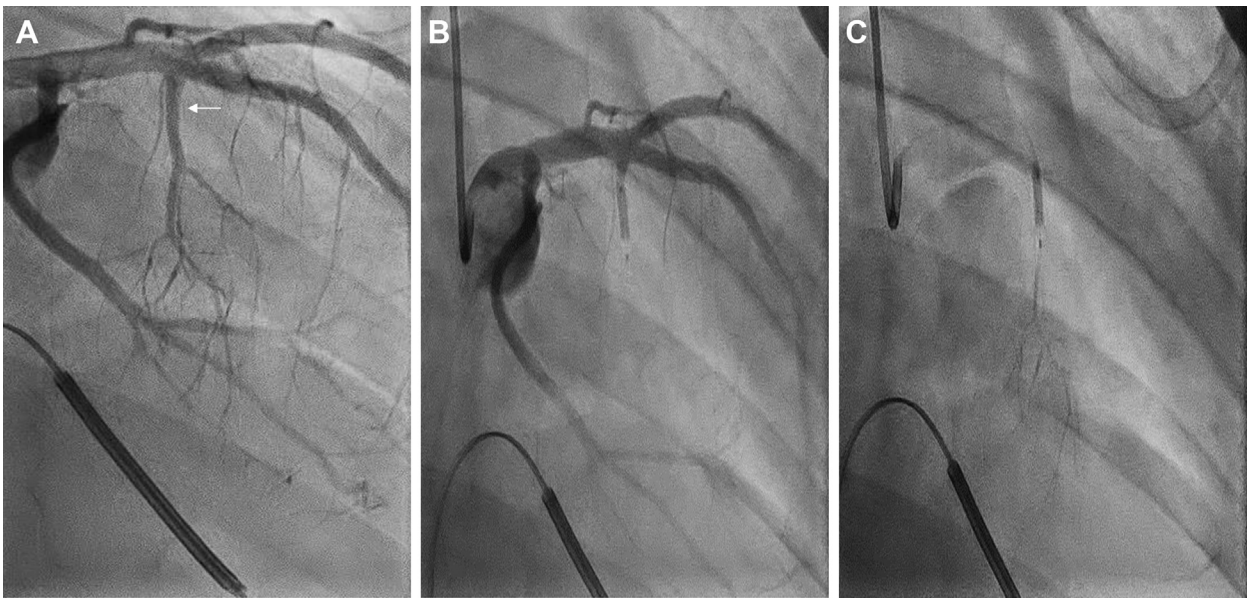
## DISCUSSION

The number of child-bearing women diagnosed with HCM is growing, due to the use of cardiac investigations and screening in HCM families. Pregnancy in women with H(O)CM bears higher risk, both for the women and for the fetuses, as there is a 40% increase of volume load and a rise of heart rate during pregnancy (1). A systematic review of 237 women and 408 pregnancies from 9 cohorts reported that the maternal mortality in pregnant patients with HCM remains low at 0.5%. However, 29% of patients developed worsening of symptoms during pregnancy, and 26% of patients had to undergo premature delivery (2). Sadly, cases of fetal death were reported (3).

Due to a thickened IVS, 37% of patients with HCM develop LVOTO at rest, defined by LVOT gradient ≥50 mm Hg (4). LVOTO with an associated

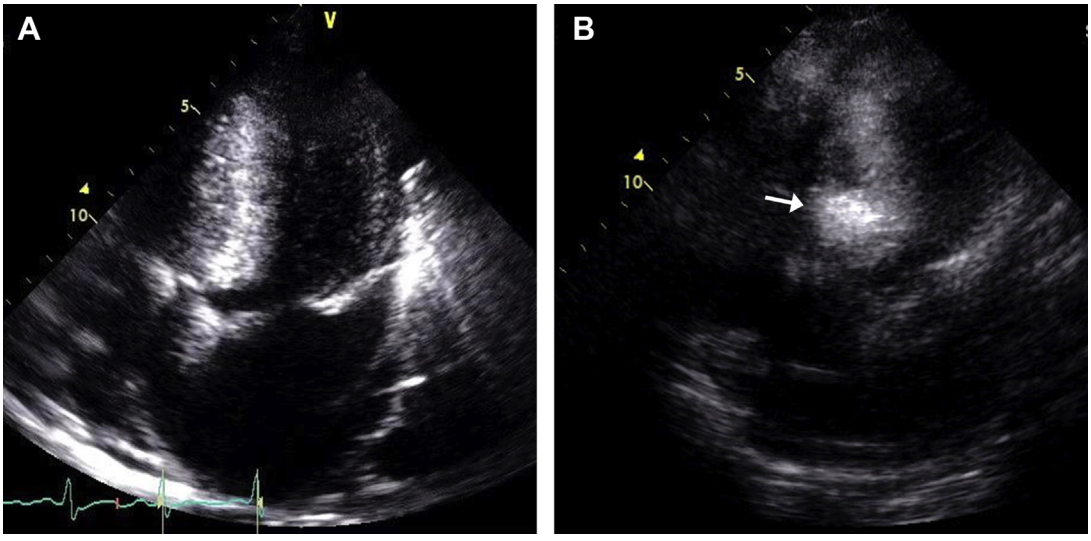
JACC: CASE REPORTS, VOL. 2, NO. 1, 2020
JANUARY 2020:139–44

**FIGURE 2 Angiogram Before and During ASA**



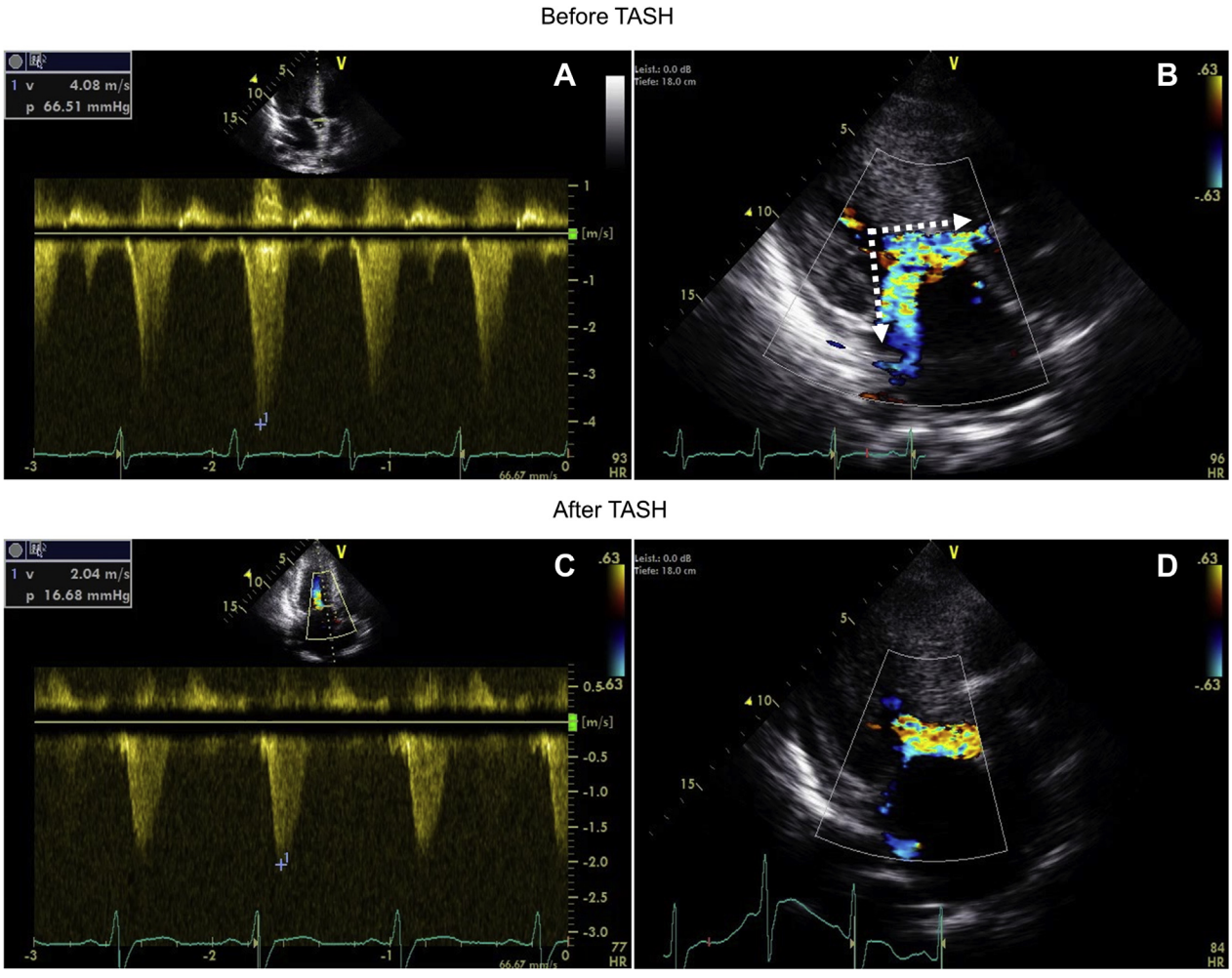

**(A)** Angiogram showing the baseline anatomy of the left coronary arteries with the large first septal perforator branch **(white arrow)**. **(B)** Angiogram demonstrating the inflated balloon in the septal branch resulting in shielding from antegrade blood flow. **(C)** To confirm the balloon position and guarantee the absence of retrograde leakage, contrast agent was injected into the first perforator branch via the lumen of the inflated over-the-wire balloon catheter. Abbreviation as in Figure 1.

**FIGURE 3 Echocardiogram Before and During ASA**




**(A)** A baseline 4-chamber view in transthoracic echocardiography. **(B)** Before the injection of alcohol, the perfusion area of the selected septal perforator was assessed in a contrast echocardiogram. The contrasted area of the basal interventricular septum **(white arrow)** suggested an optimal target for alcohol ablation. Importantly, no other region such as the free RV wall was contrasted, which could be due to collaterals. Suboptimal recordings have been tolerated in the severely dyspneic women. RV = right ventricle; other abbreviation as in Figure 1.

**FIGURE 4 Reduction of LVOT Gradient After ASA**




**(A)** Before ASA, transthoracic echocardiogram demonstrating an increased LVOT gradient of 67 mm Hg at rest, and **(B)** a hypertrophied left ventricle with SAM of the mitral valve resulting in severe mitral regurgitation. **Arrows** indicate the typical "split sign." **(C)** After ASA, the LVOT gradient has decreased to almost physiological values (16 mm Hg), and **(D)** the previously observed SAM of the mitral valve and mitral regurgitation have regressed. SAM = systolic anterior motion; TASH = transcoronary ablation of septal hypertrophy; other abbreviations as in Figure 1.

mitral regurgitation is a pathological hallmark of HOCM, and an independent predictor of adverse events, including progressive heart failure and SCD (4). According to the current recommendations, there are only a few options available for pregnant patients with symptomatic HOCM (5). In most cases, the use of β-blockers is recommended, preferably metoprolol. Second-line drugs include disopyramide, verapamil, diltiazem, and amiodaron, which are all linked to possible adverse effects on the fetus, such as maternal uterine contractions, delayed fetal development, neurologic adverse effects, and thyroid toxicity.

ASA is an established catheter-based intervention in nonpregnant patients. It especially benefits high-risk patients, who have relevant LVOTO but are contraindicated to surgical myectomy due to the risks of general anesthesia. During ASA, alcohol is injected into a suitable (often the first) septal perforator branch of the left anterior descending artery, aiming to induce a regional myocardial infarction and consecutive scaring of the IVS. It was reported that up to 92% of patients showed an immediate reduction of LVOT gradient >50% after ASA when using intraprocedural contrast-echocardiographic monitoring, which is a standard method nowadays; 94% of the

patients described improved symptoms within 3 months after ASA (6). These results are comparable to the outcomes of surgical myectomy (7).

To our knowledge, the present case is the first uncomplicated ASA performed as a rescue intervention in a pregnant woman with decompensated HOCM. Aside from our present case, there was only one other report that was published after performing this procedure in a pregnant patient, who had postprocedural complications (8). Notably, the patient in the present case had an LV septal thickness of 38 mm before ASA, in comparison with the patient in the other reported case with an LV septal thickness of 20 mm. Furthermore, the patient in the present case had received an implantable defibrillator 7 years before this event and hence we did not fear the consequences of a higher degree AV blockage, which is the major complication in up to 10% of patients with ASA.

During follow-up examinations, the patient remained asymptomatic. Due to the slight growth delay of the fetus shown by the sonography before the ASA procedure, the gynecologists and the patient decided on a cesarean delivery in analgosedation at 34 weeks of gestation. The cesarean delivery was performed without complications and the patient gave birth to a healthy infant weighing 1,990 g. Three days later, the patient was discharged. Before her discharge, long-term birth control with effective contraception was discussed to avoid unplanned pregnancy. At the 3-month follow-up examination, the LVOT gradient in the echocardiogram was <10 mm Hg, and the basal IVS thickness was reduced from 38 mm to 30 mm.

## CONCLUSIONS

The current case presents the possibility of safely performing ASA on high-risk pregnant patients, when conservative therapy is ineffective. The results of this case suggest that proactive invasive gradient reduction strategies before planned pregnancies in selected high-risk patients with HOCM could be an effective means to reduce the hazard of LVOTO deterioration during pregnancy. This option should be discussed during patient consultations.

**ADDRESS FOR CORRESPONDENCE:** Dr. Benjamin Meder, Department of Medicine III, University of Heidelberg, INF 410, 69120 Heidelberg, Germany. E-mail: Benjamin.Meder@med.uni-heidelberg.de.

## REFERENCES

**1.** Krul SP, van der Smagt JJ, van den Berg MP, Sollie KM, Pieper PG, van Spaendonck-Zwarts KY. Systematic review of pregnancy in women with inherited cardiomyopathies. Eur J Heart Fail 2011;13:584–94.

**2.** Schinkel AF. Pregnancy in women with hypertrophic cardiomyopathy. Cardiol Rev 2014;22:217–22.

**3.** Goland S, van Hagen IM, Elbaz-Greener G, et al. Pregnancy in women with hypertrophic cardiomyopathy: data from the European Society of Cardiology initiated Registry of Pregnancy and Cardiac disease (ROPAC). Eur Heart J 2017;38:2683–90.

**4.** Maron MS, Olivotto I, Zenovich AG, et al. Hypertrophic cardiomyopathy is predominantly a disease of left ventricular outflow tract obstruction. Circulation 2006;114:2232–9.

**5.** Authors/Task Force members, Elliott PM, Anastasakis A, et al. 2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy: the Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC). Eur Heart J 2014;35:2733–79.

**6.** Faber L, Seggewiss H, Gleichmann U. Percutaneous transluminal septal myocardial ablation in hypertrophic obstructive cardiomyopathy: results with respect to intraprocedural myocardial contrast echocardiography. Circulation 1998;98:2415–21.

**7.** Alam M, Dokainish H, Lakkis NM. Hypertrophic obstructive cardiomyopathy-alcohol septal ablation vs. myectomy: a meta-analysis. Eur Heart J 2009;30:1080–7.

**8.** Shaikh A, Bajwa T, Bush M, Tajik AJ. Successful alcohol septal ablation in a pregnant patient with symptomatic hypertrophic obstructive cardiomyopathy. J Cardiol Cases 2018;17:151–4.

**KEY WORDS** alcohol septal ablation, decompensation, hypertrophic obstructive cardiomyopathy, pregnancy