

# Two cases of “stone heart” with fatal outcome

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**Abstract:** The “stone heart” syndrome is a rare but often fatal complication of cardiac surgery associated with hypertrophy of the myocardium. The mechanisms behind the syndrome are not fully understood. In this case report, we describe two cases of stone heart in newborn girls. Both girls were born with congenital heart abnormalities including ventricular septum defects (VSD), hypertrophy of the myocardium and patent arterial duct (PDA), which was prenatally diagnosed. In each of the two cases, the stone heart became evident immediately after initiating cardiopulmonary bypass, and ended fatally.

**Keywords:** Stone heart; ventricular septum defects (VSD); hypertrophy; patent arterial duct (PDA)

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## Introduction

The “stone heart” syndrome is a rare and fatal complication described in neonates and adults undergoing cardiac surgery with application of cardio pulmonary bypass (1,2). Common findings in stone hearts are myocardial hypertrophy and fibrosis. The left ventricle becomes firmly contracted and in some cases presents with ventricular fibrillation (3). We describe two fatal cases of stone heart. Both cases occurred immediately after initiation of cardiopulmonary bypass.

## Case presentation

### Case 1

A girl of 870 grams was prenatally diagnosed with ventricular septum defects (VSD), hypoplastic aortic arch and a large patent arterial duct (PDA). The girl was delivered by caesarian section in gestational week 27+5 due to severe preeclampsia.

Initially the girl was intubated and put on a ventilator. She received diuretics and total parenteral nutrition. There was no need for infusion of prostaglandin.

After 2 months in the neonatal ward the weight had increased to 2,110 grams and surgery was performed. Arterial cannulation was performed using a Gore-Tex<sup>®</sup>

tube sewn to the right carotid artery and venous bicaval cannulation. Ten seconds after initiation of cardiopulmonary bypass the heart went into a fixed state of hypercontractility and stone heart was evident.

Antegrade cardioplegia with a solution set to 4 °C was attempted to relieve the heart. However, this only increased the contractility of the myocardium. It was decided to continue the operation. The corrections of the cardiac abnormalities were performed despite the edematous and contracted state of the myocardium. With each dose of cardioplegia the contraction increased, but slowly decreased afterwards.

The priming solution of 300 mL used during the operation contained 75 mL of Ringer’s lactate solution, 0.5 mL of heparin (5,000 IU/mL), 135 mL of SAG-M, 75 mL of FFP, 1.1 mL of Solu-Medrol (62.5 mg/mL), 7.3 mL of mannitol, 0.22 mL of Diural<sup>®</sup> and 6 mL of 8.4% NaHCO<sub>2</sub>.

The arterial blood gas test showed no significant deviations.

After 145 minutes the cross clamp was removed, and despite of this no pulsative contractions were evident. External pacing only achieved minor contractions. Despite two hours of reperfusion and a new run of cardiopulmonary bypass, weaning from the pump was unsuccessful due to

profuse bleeding and need of volume. For the same reason it was not possible to place the patient on extracorporeal membrane oxygenation (ECMO). The operation was finally abandoned after 8 hours. Autopsy was not performed in respect for the parents.

### Case 2

A premature girl born in gestational week 31 was diagnosed with a large perimembranous VSD and PDA. Surgery was performed at the age of 5 weeks.

When introducing cardiopulmonary bypass ventricular fibrillation occurred immediately. After unsuccessful defibrillation the heart went into a hypercontractile state. In an attempt to resolve the hypercontractile state, blood cardioplegia was initiated without any effect on the contracted myocardium.

Approximately 15 minutes after initiation of cardiopulmonary bypass the abdomen became enlarged. The liver was displaced into the mediastinum and made further access to the heart difficult. In order to enable surgery the abdomen was opened to allow for pressure relief. No signs of intra- or retroperitoneal bleeding were seen. No abnormalities in neither the descending aorta, portal vein, nor inferior vena cava were present to explain the enlarged abdomen. The inside of the right ventricle could now be inspected. The myocardium was extensively hemorrhagic. At this point the heart had gone into a more flaccid state. Blood cardioplegia was repeated, this time in a retrograde fashion. Immediately the myocardium became severely contracted. The VSD was closed using a Gore-Tex<sup>®</sup> patch. After declamping the aorta, reperfusion was commenced, but due to acidosis, hyperkalemia, no signs of cardiac recovery and an obvious non-existing peripheral circulation, surgery was stopped.

The autopsy revealed that the digestive tract was dark, swollen and ischemic from the ventricle to the rectum. The lumen contained fluid and a little blood. The psoas muscles were dark with signs of fresh bleeding. Inspection of the heart showed that the surgical corrections had gone as planned. Both the right and left side of the myocardium was a dark grey-red color, worst on the left side and in the septum. Microscopic examination of the myocardium revealed ischemic changes and fresh diffuse bleeding. The septum and posterior wall of the myocardium showed more signs of more extensive fresh bleeding with the cells being separated by focal bleeding. Microscopy of the lungs showed small alveoli and fibrosis of the parenchyma, but no

signs of inflammation.

For both cases no technical abnormalities were reported such as drops in pressure, air embolism etc. There were no anatomical abnormalities of the coronary arteries. The hypertrophies of both hearts were eccentric and not localized to the septum or apex in particular. Standard cannulation, anesthesia and medication were all undertaken without any deviations observed. Cardioplegia in both cases were first applied after development of the hypercontractile state. In neither of the cases were inotropes used during the course of treatment.

### Discussion

Two cases of stone heart in neonates have been presented. In both cases the patients had a VSD, PDA and hypertrophy of the myocardium. The literature regarding stone heart is very scarce and there are no studies published in recent years. In contrast to previous reports (1,2,4,5), development of stone heart did not occur in relation to reperfusion.

The mechanisms behind the syndrome remain not fully understood. It has been suggested that the contracted state of the stone heart can be compared to rigor mortis of striated muscles, where the contraction comes from an adenosine triphosphate (ATP)-deficiency (3,4). Animal studies of neonates have shown that hearts with lower levels of ATP are more likely to transition into stone hearts faster than hearts with normal levels of ATP (6). Since hypertrophied hearts have low resources of ATP, this may be the cause of the heart's inability to relax. Fluctuations in intracellular calcium and degradation of the sarcoplasmic reticulum can also be a cause of contraction of the heart (7). Reperfusion-induced stone heart can be caused by a calcium overload if the reenergization of the cells is rapid, and by rigor contracture if the reenergization is slow (4). Experimental studies on dogs have shown that intracellular changes come following a prolonged period of anoxic cardiac arrest which leads to myofibrillar degeneration, proportional to the time of the anoxic cardiac arrest (8). Others argue that time does not play a significant role in surgery, unlike the significance of the ultrastructure of the heart (3). This is in accordance with our two cases where the hypercontractile state became evident immediately following initiation of cardiopulmonary bypass.

Patients with congenital heart disease often have pressure or volume overloaded cardiac chambers and therefore may be more prone to myocardial ischemia (6). The use of propranolol and hypothermia has been proven to reduce

the risk of stone heart in adults and in animal studies of neonates (1,6). Intermittent stretching of the myocardium has been shown to decrease ischemic contraction during surgery on rabbits (9). However, no studies have examined the effect of these measures in neonates.

The stone heart is a rare and dreaded complication. However, despite of improved surgical techniques and refinement of cardiac pulmonary bypass it still occurs in the recent era of cardiothoracic surgery. Further research on the stone heart syndrome is necessary to identify and potentially treat those at risk.

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### Footnote

*Conflicts of Interest:* The authors have no conflicts of interest to declare.

*Informed Consent:* The parents of both the patients in this case report have given informed consent to both the writing and publication of this case report.

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