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Case report

An unusual pathway of cardiac surgery repair of aortic and mitral valve – case report

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Abstract

Prenatal diagnosis and the surgical treatment of the patient with complex aortic and mitral valve disease are rarely presented in scientific literature.

At the 30th week of gestation a 34-year-old multigravida multipara was referred by a primary care obstetrician for a detailed ultrasound examination with a suspicion of complex aortic and mitral valve disease. The patient was born with critical aortic stenosis and mitral valve insufficiency and decreased left ventricular (LV) contractility due to endocardium fibroelastosis. On the second day of life the patient underwent balloon aortic valvuloplasty in the cath lab. At 3 months of age the patient underwent surgery. Mitral valve plasty and anterior left ventricular outflow tract (LVOT) incision were performed. At the age of 9 months the patient revealed circulatory insufficiency caused by mitral stenosis and recurrent insufficiency, and was qualified for implantation of a mechanical valve in the mitral position. Intraoperatively it was noted that mitral annulus was too small to implant a mechanical valve, so a biological valve was prepared with the CMx ECM on the operating table. The implantation of a CMx biological valve custom-made on the operating table was the bridge of surgical treatment for implantation of a mechanical valve in the future. The patient successfully underwent surgery. At 3 years old, we replaced the CMx valve with a mechanical St. Jude.

This new pathway of cardiac surgery repairs in the first year of life might be an important issue for counselling parents-to-be after detection and diagnosis of prenatal congenital heart defects, such as in the presented case: abnormal aortic and mitral valve.

Key words: prenatal aortic stenosis, mitral valve insufficiency, prenatal diagnosis, balloon valvuloplasty after delivery, lyophilised extracellular matrix.

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Introduction

The development of prenatal cardiology in Poland is proven by the increasing number of prenatal cardiology centres [1]. In 20-24 pregnancies out of 1000, the development of the heart is not normal [2]. The prevalence rate of aortic valve stenosis among all neonates with heart defects is estimated at 2.5%. It is more common in male newborns (in a 4 : 1 ratio). This defect may pose an instant danger to life in tghe neonatal period and requires urgent interventional treatment. The abnormalities in the development of the aortic valve reach various stages

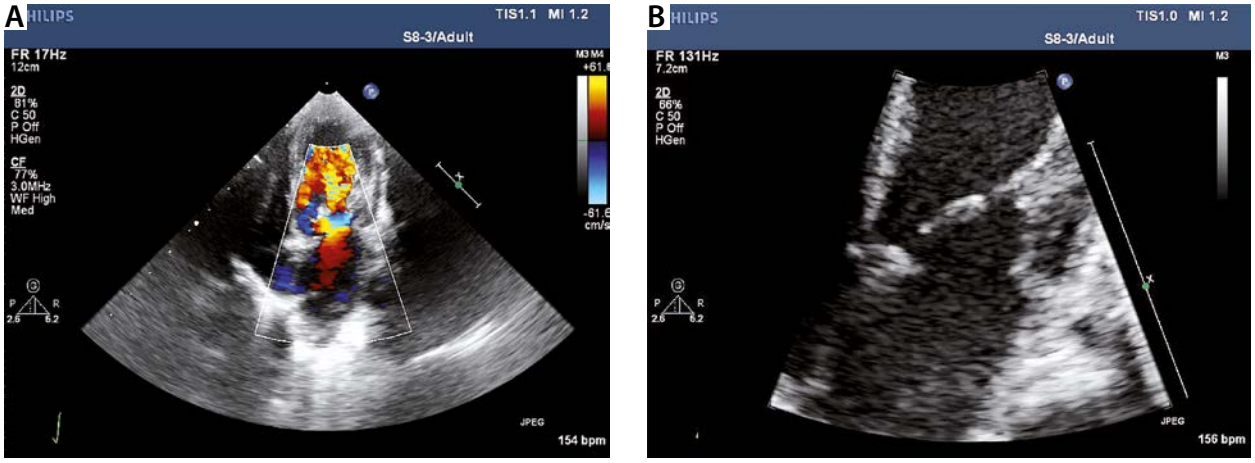

Figure 1. Echocardiographic examination before plasty of the mitral and aortic valve

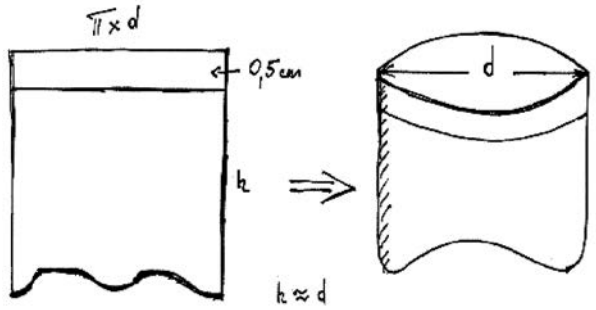

Figure 2. The draft of CMx valve creation

and thus have different effects on haemodynamics and clinical manifestation [3]. Extreme forms of valvular stenosis in the newborn accompany hypoplasia syndrome of the left heart, the presence of which is associated with ring hypoplasia (< 6 mm) [4], a decrease in the volume of the left ventricle (< 25 ml/m^2) [5], and the accompanying hypoplasia of the mitral ring (diameter less than 11 mm) [6]. Any pathology that manifests itself in the first month of a child's life is always dangerous. If the defect pertains only to the aortic valve, it most often consists of the incorrect connection of the cusps and their thickness. The valve is then built of very thick, stiff cusps, with the bonding points difficult to define. In extreme cases, it is a thick fibrous membrane with a narrow opening. Aortic annular hypoplasia of varying degrees has been associated with unformed sinuses of Valsalva.

Case report

At the 30th week of gestation a 34-year-old multigravida multipara was referred by a primary care obstetrician to a highly specialised centre for a detailed ultrasound examination with a suspicion of complex aortic and mitral valve disease. The patient was born (38 hbd, Apgar score of 9 points) by vaginal delivery with critical aortic stenosis and mitral valve insufficiency and decreased left ventricle contractility due to endocardium fibroelastosis. On the second day of life the patient underwent balloon aortic valvuloplasty in the cath lab. The gradient across the aortic valve was gradually increased up to 40 mm Hg, and in the third month of life he underwent a surgical procedure (Figure 1). During an open heart surgery a parachute type of mitral valve was revealed and the single papillary muscle was split into two parts with separation of tendinous chordae of the anterior leaflet. The anterior commissure of functional bicuspid aortic valve was incised to the annulus and gentle shaving of the thickened cusps was done. Anterior incision of the left ventricular outflow tract (LVOT) muscle was done. The small interatrial foramen was left opened for decompressing of the left atrium if needed. Postoperative echocardiographic assessment revealed mild mitral regurgitation (II degree), and the transvalvular aortic mid gradient reached 25 mm Hg.

Six months later, at the age of 9 months, the patient revealed circulatory insufficiency caused by mitral stenosis and recurrent insufficiency. The aortic valve function was acceptable with an echo gradient of 25 mm Hg. The patient was qualified to implantation of a mechanical valve in the mitral position. Intraoperatively it was noted that the mitral annulus was too small (12 mm) to implant a mechanical valve. Based on our previous experience regarding implantation of a biological valve prepared from ECM CMx on the operating table, we decided to fashion a tube-like valve and implant it into the mitral position. The valve was sewn from a 4 × 7 cm sheet of extracellular matrix (ECM) like a tube 16 mm in diameter. The height of this valve was 20 mm (125% of the valve diameter). There were two anchoring points at the level of native papillary muscles enforced with Dacron pledgets. The proximal annular anastomosis was performed applying 5.0 Prolene suture. To reinforce this junction a 5 mm cuff of ECM folding was created. The postoperative course was uneventful, and the patient was discharged home 2 weeks after operation. Aspirin as an antiplatelet drug was administered during follow-up for 3 months (Figures 2-6).

He was in continuous outpatient follow-up. The consecutive echo assessment revealed increased mitral stenosis up 12 mm in the annular dimension and 4 mm in transvalvular flow. The leaflets were thickened. The stenotic flow histogram was revealed the velocity up to 2.7 m/s and its mild insufficiency. Aortic valve insufficiency was mild, and trivial stenosis was noted (Figure 7).

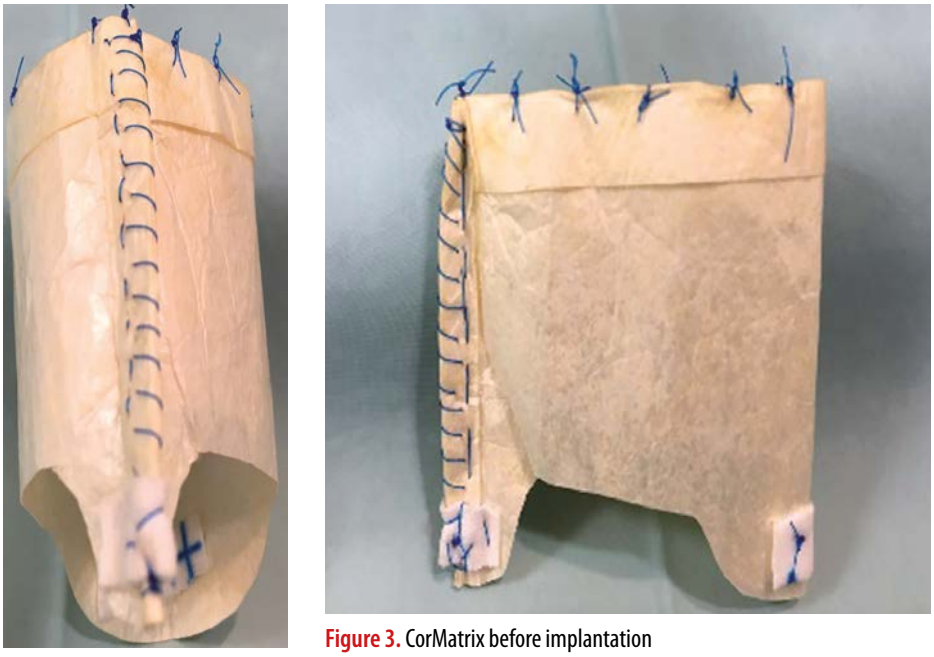

Figure 3. CorMatrix before implantation

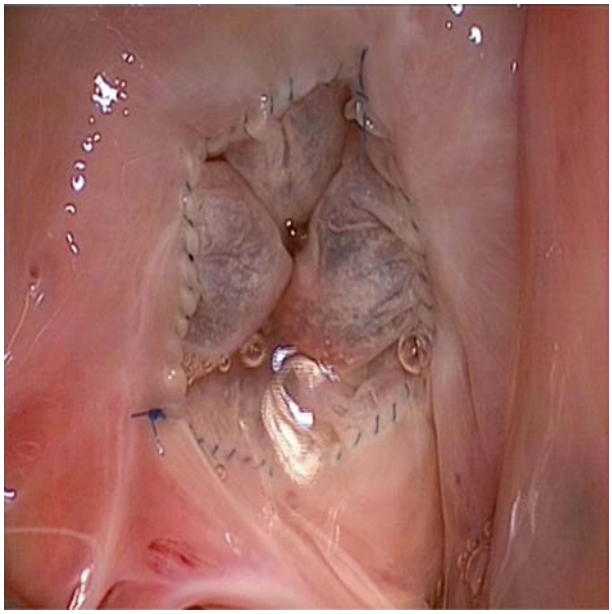

Figure 4. CorMatrix valve – intraoperative view

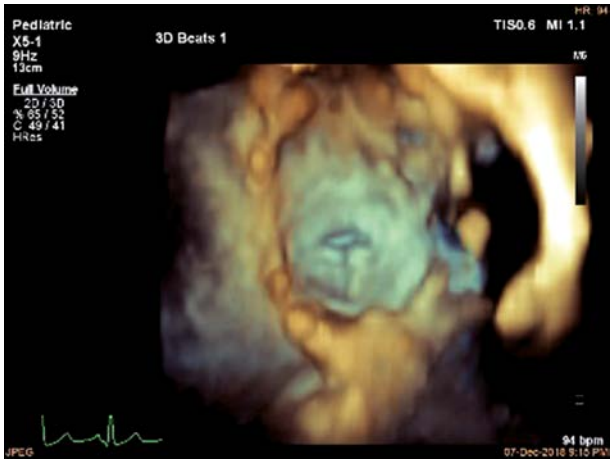

Figure 5. 3D – CorMatrix valve in mitral position

The patient successfully underwent the subsequent surgery. We replaced the CMx valve with a mechanical St. Jude 15 mm in diameter (Figure 8). Echo examination showed decrease of the left atrium and good MV functioning (Figures 9, 10).

Discussion

Reconstructive cardiac surgery is a challenging performance for congenital heart diseases, and application of a patient's own tissue is frequently insufficient for optimal effects. A lot of substitutes (either biological and prosthetic) that could be used as a patch, i.e. autologous pericardium (fresh or glutaraldehyde – fixed), xenopericardium (bovine or equine), polytetrafluoroethylene – PTFE, or full valved conduits (freeze homografts or xenografts) have not met expectations. Mid-term and especially long-term outcomes are still suboptimal. Autologous or cross-linked xenopericardium is susceptible to degenerative processes including fibrosis, scar formation, calcification, and (extremely harmful to patients) the lack of potential growth of implants. Biological valves cannot always meet demands especially regarding their size and availability. The paediatric population has particular needs because bioprostheses, homografts, and xenografts are susceptible to accelerated degeneration. There is an urgent need to find materials for use in cardiovascular surgery to overcome the listed limitations. CorMatrix-SIS biological scaffold was proved to be a promising, innovative material in experimental and clinical cardiovascular surgery. Clinical results support the notion that this material may possesses many features of an 'ideal' biological scaffold,

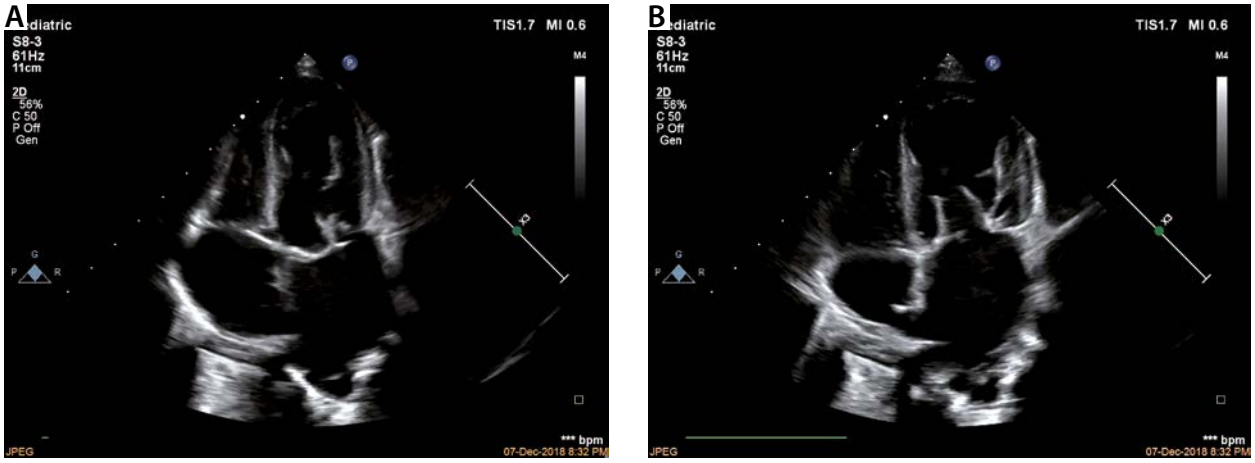

Figure 6. CorMatrix valve in mitral position

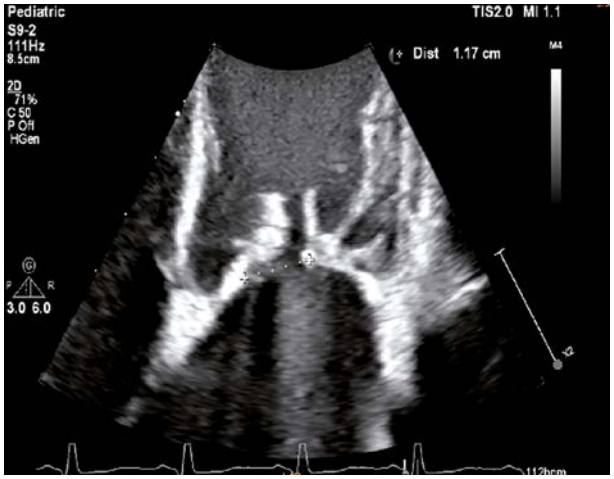

Figure 7. CorMatrix valve in mitral position prior to removal

such as like strength and durability, feasibility in clinical applications, and simplicity in creation on the operating table. ECM is a material derived from different tissues, which is supposed to provide an interim bioscaffold that enables the patient's own stem cells and surrounding tissue cells to migrate, repopulate, and repair damaged tissue by replacing the degrading bioscaffold over time. The remodelling of extracellular matrix subintestinal submucosa (ECM-SIS) scaffold has been reported in animal studies and in human clinical applications [7]. However, the long-term outcomes and follow-up of patients who have undergone this kind of surgery is still unknown.

The most challenging issue is the reconstruction of the mitral valve in the paediatric population. Mechanical valves are not intended for use in neonates and toddlers. The smallest size available on the market is 15 mm in diameter, and it is often too large to be implanted in neonates. CorMatrix is a relatively new construct made of extracellular matrix, which has previously been used to repair pulmonic and tricuspid valves. The other possibilities of replacement, including the specially prepared melody valve in the mitral position or inverted pulmonary valve in prosthesis tube, have not met expectations.

Conclusions

This new pathway of cardiac surgery repairs in the first year of life might be an important issue for counselling parents-to-be after detection and diagnosis of prenatal congenital heart defects, like in the presented case: abnormal aortic and mitral valve.

Conflict of interest

The authors declare no conflict of interest.

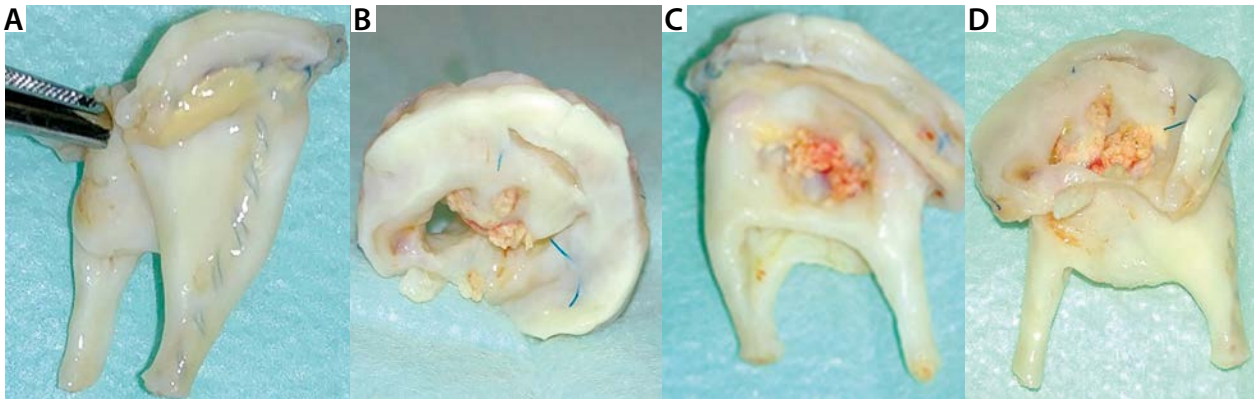

Figure 8. Removed CorMatrix valve from mitral position – visible calcium changes on the valve leaflets

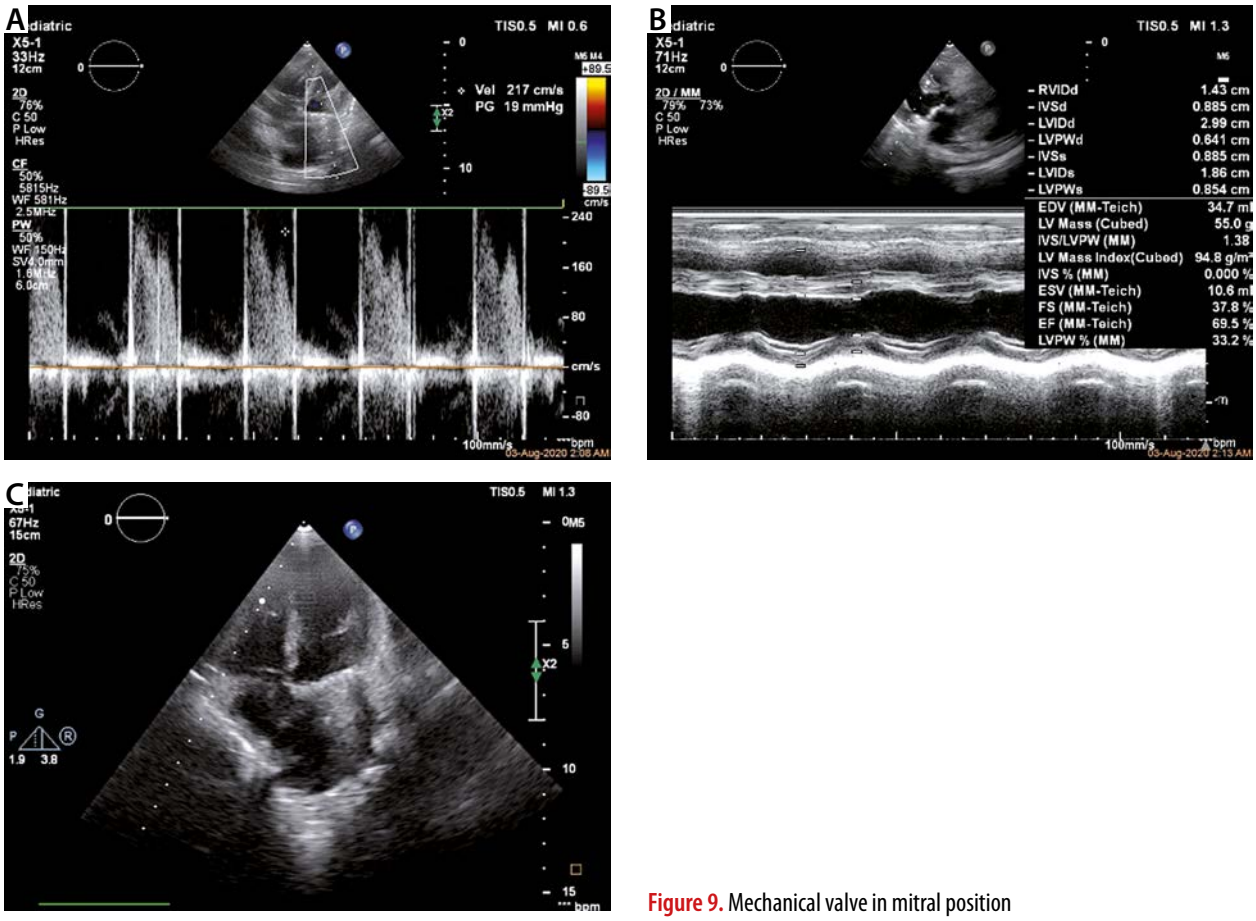

Figure 9. Mechanical valve in mitral position

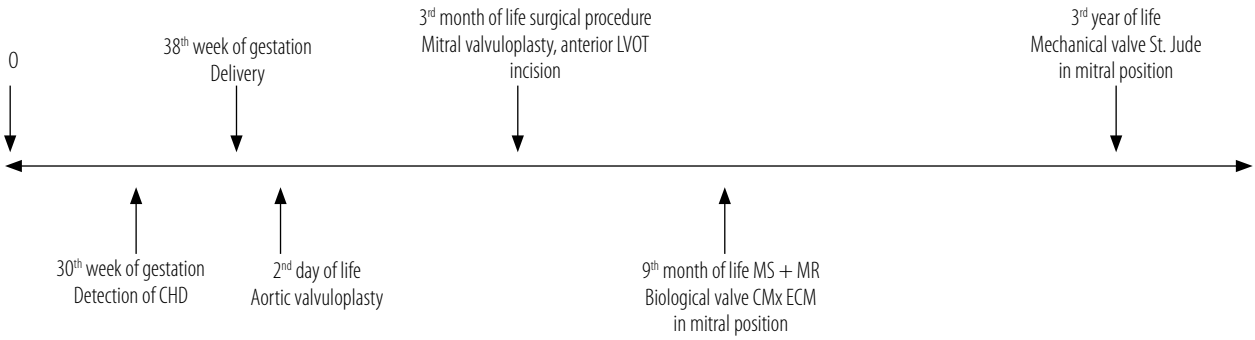

Figure 10. Steps of the consecutive procedures – lifeline

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