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Multidisciplinary Support for Pregnant and Parturient with Congenital Heart Disease

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Abstract. Adults with corrected or uncorrected congenital heart defects form a separate cohort of patients – GUCH (grown-up with congenital heart). GUCH patients usually have a certain degree of heart failure, arrhythmias, an increased risk of infective endocarditis; some of them require additional heart surgery, with an increased risk of developing sudden cardiac death syndrome. Experts from the European Society of Cardiologists are considering questions and medical support for pregnancy, childbirth and postpartum period in women from a GUCH cohort. In accordance with the ESC recommendations, a multidisciplinary team consisting of specialists from National Amosov Institute of Cardiovascular Surgery and the Institute of Pediatrics, Obstetrics and Gynecology of NAMS of Ukraine, was created in Ukraine in 2013.

Objectives. Reducing maternal and infant mortality and disability, as well as providing optimal management and delivery strategy for pregnant women with congenital cardiac pathology.

Materials and methods. Counselling assistance was provided to 2 815 pregnant women with cardiac pathology, 116 women underwent inpatient treatment since 2013. The group of GUCH patients consisted of 64 women, 61 of them were pregnant, 3 were parturient. 52 patients had structural CHD, 12 women suffered from congenital anomalies of the cardiac conduction system. There were patients with a corrected CHD and satisfactory result (n=4), with CHD after palliation or significant residual shunts of the defect (n=13), and GUCH pregnant with uncorrected CHD (n=35). Some inpatient women received non-surgical treatment (n=11, 17%). In 53 women (50 pregnant and 3 parturient), who underwent any type of surgery, 42 cardiac and vascular interventions, 18 caesarean sections were performed. Cardiac surgeries included endovascular interventions (n=22), operations with CPB (n=19), one intervention on the femoral artery. There were 23 elective operations, and 19 urgent or emergency cases.

Results. One maternal and two perinatal losses were observed in a group of pregnant women (parturient) who underwent emergency intervention with CPB. There was no more maternal loss or perinatal loss in all other cases (n=47), pregnancies finished either on a term delivery or on a Caesarean section without complications with high functional indicators in newborns by the Apgar scale. The current level of cross-sectoral specialized care for GUCH-pregnant with special needs greatly reduces the risk of mortality and disability in the mother and child. **Conclusions.** The first line of defence of GUCH-women of childbearing age involves primary prevention of major cardiac events, namely preconceptional counselling at specialized expert centres, and their pregravid preparation, multidisciplinary support of such women throughout pregnancy, delivery and postpartum period.
Keywords: *pregnancy, GUCH, multidisciplinary care.*

Congenital heart defect (CHD) is an abnormal structure and function of the heart and/or great vessel that exists at birth. Congenital heart defects are formed as a result of impaired embryogenesis in the period from Week 2 to Week 8 of gestation upon angiogenetic accumulation of cells at the site of cardiac germination (Day 18 + 1 of gestation) to complete formation of the aortic valve cusps and sinuses of Valsalva (approximately Day 55 of gestation) [1]. The epidemiology of patients with CHD is 7 to 9 cases per 1,000 live births [2].

Today, according to the classification offered by the World Cardiac Surgical Society, there are more than 160 types of CHD and more than 210 techniques of cardiac surgery for them [3].

In the current cardiac surgical era, approximately 85% of CHD infants reach adulthood, with the number of patients after heart surgery in the childhood or non-corrected CHD 2800 per 1 million [4]. According to the literature, the overall survival of patients after CHD correction up to 40 years reaches 95%, up to 50 years – 92%, up to 60 years – 88% [5].

Adults with corrected or uncorrected CHD form a separate cohort of patients – GUCH (grown-up with congenital heart).

The quality of life of patients after heart surgery in the childhood is a controversial issue. On the one hand, according to literary sources [6], such children are more likely to be behind at school, more likely to miss school, more likely to have allergic manifestations and neurological retardation. But in adulthood, patients after heart surgery for CHD show a fairly active social position in society. As evidenced by the study of E.H.M. van Rijen [7], most of these patients have families, almost one in three have children. And for many of them, having an operated heart is an additional motivation for a more active lifestyle, including the function of motherhood.

GUCH patients typically have some degree of heart failure, due to a number of factors – such as residual septal defects, pulmonary hypertension, uncorrected CHD

components, and palliative interventions. Arrhythmias are a major cause of hospitalization of adults with CHD. Sudden cardiac death syndrome is another characteristic complication of the remote period in this cohort of patients [8]. GUCH patients are at increased risk of developing infectious endocarditis, some of them requiring heart (lung) transplantation, implantation of mechanical cardiac support devices, surgery and interventions for residual CHD components. Experts from the European Society of Cardiologists consider separately the issues of medical support for the pregnancy, childbirth and postpartum period in women from the GUCH cohort [9].

According to the world literature [10], pregnancy carrying (more than 20 weeks of gestation) in women with CHD do not exceed 90% for simple CHD and 48-50% for complex ones. The prognosis for pregnancy is especially unfavourable with the following CHD: Ebstein anomaly, corrected transposition of the great vessels (TGV), tetralogy of Fallot, partial atrioventricular septal defect, cyanotic CHD, hemodynamically corrected CHD (Fontan circulation). In addition, such women have a significant risk of foetal hypotrophy, embryo-and fetopathies [11].

Today, a considerable number of women of childbearing age with operated heart or “small” forms of the CHD, as well as women after palliative surgeries or those who at one time underwent hemodynamic correction of complex CHD, live in Ukraine. Of these, about 700-1 000 per year reach the childbearing age, with an annual increase of 200-300 persons. According to estimates, up to 12,000 women with heart surgery are currently living in Ukraine. All of these women require mandatory pre-conceptual counselling and pregravidary preparation. Only such a modern approach and multidisciplinary support for pregnant women with cardiac pathology minimize the risks for both the mother and child.

In accordance with the recommendations of the European Society of Cardiologists [10], a multi-disciplinary team consisting of specialists from two academic institutions – the State Institution “National Amosov Institute of Cardiovascular Surgery of the National Academy of Medical Sciences of Ukraine” and the State Institution “Institute of Pediatrics, Obstetrics and Gynecology of National Academy of Medical Sciences of Ukraine”, was created in Ukraine in 2013.

The objective of our cooperation is to reduce maternal and infant mortality rates and the disability of pregnant women (parturients) and new-borns.

Materials and methods

From December 2013 to February 2019, 2815 pregnant women required specialized multidisciplinary care, who were referred to the Counselling Polyclinic of National Amosov Institute of Cardiovascular Surgery. After conducting expert cardio-screening and stratification of the cardiovascular maternal risk, we separated a group of pregnant women with “special” needs, who were under our observation together with the specialists of the Institute of Paediatrics, Obstetrics and

Gynaecology during the entire pregnancy, childbirth and postpartum period. In the presence of a high cardiovascular risk, the patients were hospitalized at National Amosov Institute of Cardiovascular Surgery for the purpose of medical therapy, cardiac surgery, or delivery. During this period, 116 women required hospitalization.

The group of GUCH patients consisted of 64 women, of whom 61 were pregnant and 3 women were parturient. 52 patients had structural CHDs, 12 women were with congenital malformations of the cardiac conduction system. Abnormal development of the cardiac conduction system were represented by congenital atrioventricular blockade (n = 9), the course of which was complicated during pregnancy, and supraventricular tachycardia in the form of WPW syndrome (n = 3). In the subgroup of pregnant women with congenital heart rhythm disorders, 11 interventions were carried out – 7 implantations of a cardiac pacemaker (PM), 1 placement of temporary PM and 3 radiofrequencies of arrhythmia foci.

The spectrum of structural congenital pathologies of the heart and great vessels was quite wide: it was represented by 15 different types of CHDs. For ease of presentation, we divided them into the following groups: a group of defects accompanied with stenosis of the left heart compartments (Table 1), "cyanotic" CHDs (Table 2), CHDs with increased pulmonary blood flow and high hypertension in the pulmonary artery (Table 3).

In addition, a cohort of GUCH pregnant women included two patients with Marfan syndrome and aortopathy.

Table 1

Group of GUCH pregnant (parturient) women with the obstruction of the left heart compartments

Pathology	Number
Aortic coarctation	7
Congenital aortic stenosis, which is usually accompanied with post-stenotic aneurysm	19
Subaortic stenosis	3
HCM, obstructive form	3
Total	32

Note. * HCM – hypertrophic cardiomyopathy.

Pregnant (parturient) women hospitalised at the Polyclinic of National Amosov Institute of Cardiovascular Surgery included patients with radically corrected CHD and a satisfactory correction result (n = 4), with CHD palliation surgery or with significant residual components of the defect (n = 13), and GUCH-pregnant women with uncorrected CHD (n = 35). The subgroup of pregnant women after radical CHD correction with a satisfactory result included patients with the following

interventions: condition after correction of sub-aortic stenosis, aortic coarctation, the Ross-Konno procedure and PM implantation (n = 1), condition after arterial switch procedure with transposition of the great vessels (n = 1), condition after radical correction of the complete form of atrioventricular septal defect (n = 1), condition after radical correction of Taussig-Bing anomaly (n = 1). The subgroup of pregnant women with CHD following palliation surgery or with unsatisfactory results consisted of patients with the following diagnoses: condition after correction of sub-aortic stenosis, presence of patent ductus arteriosus (PDA) with pulmonary arterial hypertension (PAH) (n = 1), condition after mitral valve replacement due to congenital mitral insufficiency (n = 1), prosthetic thrombosis (n = 1), condition after mitral valve replacement due to its congenital insufficiency, severe mitral insufficiency (n = 1), corrected transposition of the great vessels, condition after narrowing of the pulmonary artery (PA) (n = 1), tricuspid atresia, condition after Glenn procedure (n = 1), pulmonary atresia, type II, tricuspid atresia, condition after Fontan procedure (n = 1), single ventricle, condition after PA narrowing, elimination of aortic coarctation (n = 1), condition after elimination of subaortic stenosis , relapse (n = 1), combined aortic defect with stenosis predominance, condition after aortic valvulotomy (n = 1), condition after elimination of aortic coarctation, relapse (n = 1), HCM, condition after Morrow procedure, relapse (n = 1), condition after aortic valve replacement, restenosis due to pannus formation (n = 1), condition after radical correction of double-outlet right ventricle, trifascicular atrioventricular block (n = 1).

Table 2. Group of GUCH pregnant (parturient) women with “cyanotic” CHD

Pathology	Number
Tetralogy of Fallot	3
Pulmonary stenosis	3
DORV	1
TGA	1
Corrected TGA	1
Single ventricle (TA, PA)	3
Ebstein anomaly	1
Total	13

Note. *DORV – double-outlet right ventricle, TGA – transposition of the great arteries, cTGA – corrected transposition of the great arteries, TA – tricuspid atresia, PA – pulmonary atresia.

The following diagnoses were reported in patients with **uncorrected** CHD: congenital aortic valve stenosis with aortopathy (n = 16), aortic coarctation (n = 6), pulmonary stenosis (n = 3), tetralogy of Fallot (n = 2), subaortic stenosis (n = 2), hypertrophic cardiomyopathy, obstructive form (n = 2), aortic aneurysm in Marfan syndrome (n = 2), Ebstein anomaly (n = 1), PDA with PAH (n = 1).

Table 3

Group of GUCH pregnant (parturient) women with increased pulmonary blood flow and PDA

Pathology	Number
Patent ductus arteriosus	2
Congenital mitral insufficiency	2
Atrioventricular septal defect	1
Total	5

Cardiovascular risk stratification was performed according to three scales: the WHO modified scale (mWHO) and ZAHARA and CarPreg scales. According to the ZAHARA scale, pregnant women scored (graded) from 0.75 points (7.5% probability of cardiac complications) to 6.75 points (70% probability of cardiac complications), with an average of 3.0 points. According to the CarPreg scale, patients scored from 0 to 2 points (risk of cardiovascular complications – 75%), with an average of 1.5 points. According to the modified WHO scale, the distribution is shown in Fig. 1.

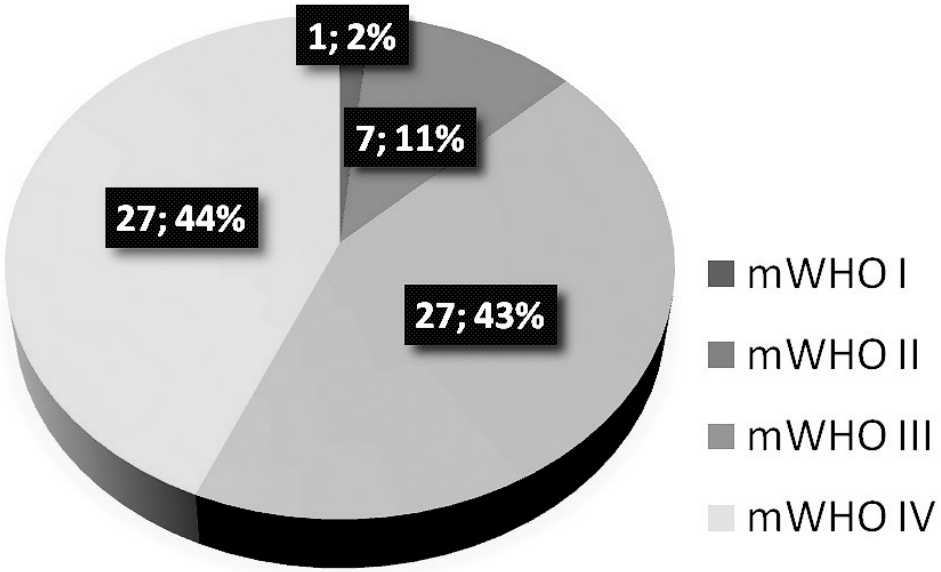

Fig. 1. Distribution of pregnant women according to the modified WHO scale of cardiovascular risk

After examination and multidisciplinary expert consultation, we determined the tactics of pregnancy and childbirth care in each case, depending on the class of the cardiovascular and perinatal risk, specific clinical situation and duration of pregnancy.

All patients were divided into the following groups according to the type of intervention: conservative, cardiac and great vessels intervention groups, caesarean section, combined surgery (cardiac surgery with caesarean section) (Fig. 2).

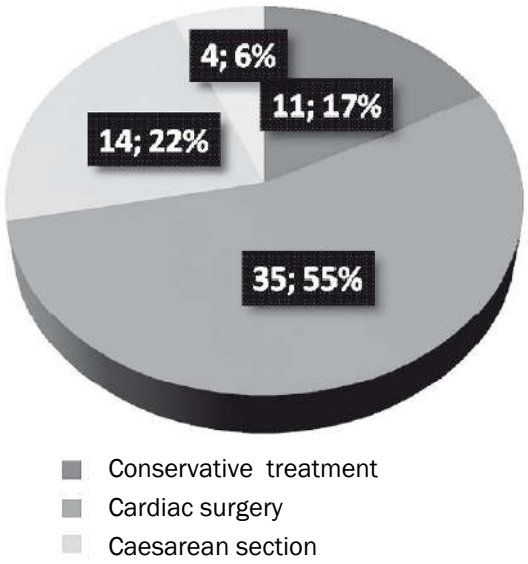

Fig. 2. Distribution of GUCH-pregnant (parturient) women depending on the chosen tactics of treatment and pregnancy and childbirth care

Of the 53 pregnant women and parturient women (50 pregnant women and 3 parturient women) who underwent surgical interventions in the clinic of National Amosov Institute of Cardiovascular Surgery, 14 underwent caesarean section, 35 patients - cardiac surgery, and in 4 cardiac surgery and caesarean section were performed. In 53 women, who underwent any type of surgery, 42 cardiac and vascular interventions, 18 caesarean sections were performed. Cardiac surgeries included endovascular interventions (n=22), operations with CPB (n=19), one closed intervention on the femoral artery. There were 23 elective operations, and 19 urgent or emergency cases of cardiac and vascular interventions.

Three parturient women underwent 1 operation with CPB, 1 hybrid intervention (endovascular + with CPB), 1 endovascular intervention and closed intervention on the femoral artery, with two of the three women being operated in an emergency procedure.

Accordingly, in pregnant women (n = 50) 17 operations with CPB and 20 endovascular operations were performed.

Types of cardiac surgery with CPB and main characteristics of patients are given in Table 4.

Table 4. Cardiac surgery with CPB in GUCH pregnant (parturient) women

No.	Diagnosis	Intervention	Urgent/ elective	Term of Pregnancy
1	AAD type A, Marfan syndrome	Bentall procedure	urgent	parturient
2	Tetralogy of Fallot	TCTOF	urgent	Week 37
3	Condition after MVR, prosthesis thrombosis	rMVR	urgent	Week 22
4	Ascending aortic aneurysm, aortic insufficiency, Marfan syndrome	Bentall procedure	elective	Week 19
5	Bicuspid AV, CAD with stenosis predominance, condition after ventricular fibrillation, CRP	AVR	urgent	Week 27
6	Bicuspid AV, CAD with stenosis predominance, ascending aortic aneurysm	AVR with mini-sternotomy + ascending aortic bandage	elective	Week 21
7	Bicuspid AV, CAD with stenosis predominance, ascending aortic aneurysm	Robicsek procedure	elective	Week 20
8	Bicuspid AV, CAD with stenosis predominance, ascending aortic aneurysm	AVR	elective	Week 18
9	AAD, type A. Aortic coarctation. Condition after caesarean section	Endovascular coarctation repair + supracoronary ascending aortic replacement	urgent	parturient
10	Discrete sub-aortic stenosis with severe symptoms	Resection of fibromuscular diaphragm	urgent	Week 18
11	Bicuspid AV, CAD with stenosis predominance, ascending aortic aneurysm	AVR	elective	Week 18
12	Common ventricle. Subaortic stenosis. Condition after PA narrowing, PDA closure, elimination of aortic coarctation	Atrioseptectomy. Bi-Di Glenn	emergency	Week 8
13	Bicuspid AV, CAD with stenosis predominance, ascending aortic aneurysm	AVR with mini-sternotomy	elective	Week 22
14	Bicuspid AV, CAD with stenosis predominance, ascending aortic aneurysm	AVR with mini-sternotomy	elective	Week 20
15	Bicuspid AV, CAD with stenosis predominance, ascending aortic aneurysm	AVR with mini-sternotomy	elective	Week 20
16	Congenital MV defect. Condition after MV replacement. Severe mitral insufficiency.	rMVR + TV replacement	elective	Week 21
17	Bicuspid AV, CAD with stenosis predominance, ascending aortic aneurysm	AVR with mini-sternotomy	elective	Week 19
18	Bicuspid AV, CAD with stenosis predominance, ascending aortic aneurysm	AVR with mini-sternotomy + implantation of a cardiac pacemaker	elective	Week 20
19	Bicuspid AV, CAD with stenosis predominance, ascending aortic aneurysm	AVR with mini-sternotomy + ascending aortic bandage	elective	Week

Note. *AV – aortic valve, AAD – acute aortic dissection, CAD – combined aortic defect, AVR – aortic valve replacement, TCTOF – total correction of tetralogy of Fallot, redo-MVR – repeated mitral valve replacement, CPR – cardiopulmonary resuscitation.

The decisions about elective cardiac surgery were made by all members of the pregnancy heart team, noting the existing cardiovascular and antenatal risks for pregnancy prolongation and deliveries, provided there was an uncorrected heart defect as well as the clinical condition of the patient and foetus. Considering the wide experience of specialists of National Amosov Institute of Cardiovascular Surgery in providing the cardiac surgery and postoperative management of patients, highly professional care of colleagues-obstetricians-gynaecologists, on the one hand, and a significant risks for preterm birth at Week 25-26 on the other hand, the following management tactics of high cardiovascular risk pregnancy was determined: (1) observation and conservative therapy in the first trimester – (2) elective cardiac surgery at Weeks 18-24 of pregnancy – (3) prolongation of pregnancy up to Weeks 36-38 with further delivery and care by a multidisciplinary team. All elective cardiac surgeries were performed with special perfusion conditions – normothermic (moderately hypothermic) perfusion with the specific of the perfusate composition, elevated parameters of the perfusion volumetric rate, blood pressure, etc. At all CPB stages, the vital signs of the foetus were monitored according to the original method [12].

Particular attention was paid by the multidisciplinary team to pregnant women in an urgent condition who underwent urgent or emergency interventions, despite the term of pregnancy and condition of the foetus. There were 19 patients with 23 interventions (Fig. 3).

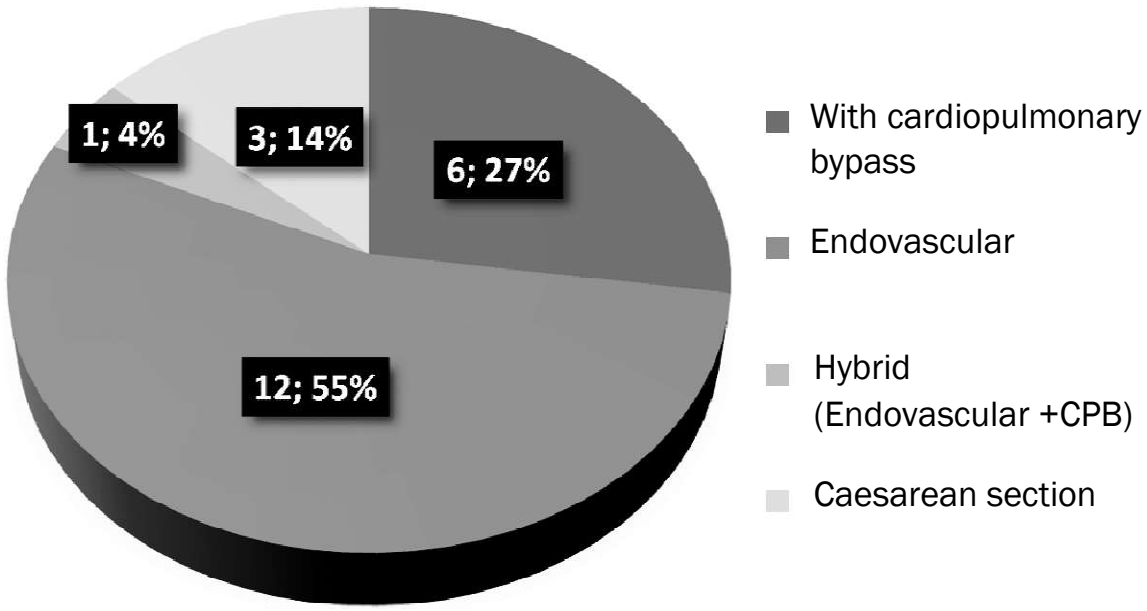

Fig. 3. Emergency and urgent interventions in pregnant (parturient) women

Endovascular interventions are mainly electrophysiological procedures for pacemaker implantation (n = 6) and urgent radiofrequency ablation (n = 2), as well as dilatation of critical pulmonary artery stenosis (n = 1) and endovascular dilation of the aortic coarctation site with endograft placement (n = 3). Endovascular interventions were performed at Weeks 15-33 of gestation, as well as in one parturient woman.

In the group of emergency and urgent interventions, one hybrid operation was performed in the parturient woman with type A AAD, which developed on Day 3 after caesarean section in the patient with moderate aortic coarctation receiving three-component antihypertensive therapy.

Emergency and urgent procedures with cardiopulmonary bypass included:

- Re-do mitral valve replacement in connection with acute prosthesis thrombosis at week 22 of gestation;

- Bentall-de-Bone procedure in a woman with type A AAD with aortopathy and Marfan syndrome at Week 40 of gestation after emergency caesarean section;

- emergency aortic valve replacement in a pregnant woman with critical congenital AV stenosis, ventricular fibrillation and cardiopulmonary resuscitation at Week 27 of gestation;

- emergency correction of tetralogy of Fallot immediately after caesarean section, spasm of the right ventricular outflow tract, cardiac arrest and cardiopulmonary resuscitation;

- Robicsek procedure (AVR + ascending aortic wall reinforcement) in a pregnant woman with congenital critical stenosis of the aortic valve (peak gradient 121 mmHg) and post-stenotic aneurysm of the ascending aorta (52 mm) due to the severe symptoms and heart failure at Week 20 of gestation;

- resection of the fibromuscular sub-aortic diaphragm and symptomatic sub-aortic stenosis (LV peak gradient 115 mmHg) in a pregnant woman at Week 18 of gestation.

Results

Immediate maternal and antenatal, as well as perinatal outcomes, depended mostly on the initial clinical condition of the pregnant (parturient) woman, the type of intervention, and the presence of an urgent condition (Table 5).

Table 5. Results of multidisciplinary support of GUCH-pregnant (parturient) women depending on the type and urgency of cardiac surgery

Intervention type	Number	Early maternal mortality	Late maternal mortality	Perinatal losses
Elective without CPB ("closed" + endovascular)	11	-	-	-
Urgent without CPB ("closed" + endovascular)	12	-	-	-
Elective with CPB	12	-	-	–
Urgent with CPB	7	1*	–	2**
Total	**42**	**1**	**0**	**2**

Note. *Pregnant, Week 40. Acute aortic dissection, Marfan syndrome. Caesarean section + Bentall procedure. Exitus on Day 8 before discharge from the hospital.

**Pregnant, Week 26. Critical aortic valve stenosis, peak aortic valve gradient of 120 mmHg, ventricular fibrillation, resuscitation, emergency aortic valve replacement. Antenatal foetal death on Day 3 after surgery

** Pregnant, Week 22. MV prosthesis thrombosis. Emergency re-do MVR. Antenatal foetal death at Week 3 after surgery.

One maternal and two foetal losses were observed in the group of pregnant women (parturient women) underwent emergency intervention with CPB, and the reasons for such consequences are quite clear.

There were no more maternal or foetal losses. In all other operated women (n = 47), the pregnancy ended either with term delivery or by caesarean section at the Institute of Paediatrics, Obstetrics and Gynaecology or National Amosov Institute of Cardiovascular Surgery without complications, with high functional parameters of new-borns on the Apgar scale.

Conclusions

1. The current level of multidisciplinary specialized care for GUCH pregnant women with "special needs" significantly reduces the risk of maternal and child mortality and disability.

2. The first line of care for GUCH-women of childbearing age involves primary prevention of major cardiac events, namely preconceptional counselling at specialized expert centres, and their pregravidary preparation, including cardiac surgeries, with further multidisciplinary support of such women throughout pregnancy, delivery and postpartum period.

3. Optimal support of pregnant women with congenital cardiac pathology requires multidisciplinary interaction of expert centres.

4. Pregnant women from the risk class III-IV modified WHO scale should be referred to the Institutes of the NAMS system of Ukraine (Institute of Pediatrics, Obstetrics and Gynecology and National Amosov Institute of Cardiovascular Surgery)

for the purpose of providing expert cardiology, cardiac surgical, obstetric and gynaecological advice.

5. In the case of emergency in pregnant woman with a high-risk cardio-pathology, urgent cardiac surgery provides an opportunity to preserve the life of the mother, although it increases the antenatal risks for the foetus.

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