

# COMPARISON OF THE CLINICAL STATUS AND THE EFFECTS OF TREATMENT OF NEWBORNS WITH INTERRUPTED AORTIC ARCH DIAGNOSED PRENATALLY AND POSTNATALLY IN THE "POLISH MOTHER'S MEMORIAL" INSTITUTE IN LODZ IN THE YEARS 2003-2012



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PRENAT CARDIO. 2014 JUN;4(2):11-19  
DOI 10.12847/06142

## Abstract

**Material and Methods:** A retrospective analysis of 41 patients with congenital heart defect IAA from ICZMP in Lodz in the years 2003-2012, including 15 (36.6%) after prenatal diagnosis.

**Results:** An average of 4 newborns with IAA were operated annually, in the years 2003 -2009 postnatal diagnosis dominated, from 2010 prenatal diagnosis. In the group,  $n = 15$  fetuses with IAA, heart size was normal: Avg  $0.33 + / -0.05$  HA / CA. Cardiovascular endurance in the CVPS : 8 - 10 points Avg.  $9.56 + / -0.73$ . Infants in the "prenatal" group were often delivered by CS (64.3%), and infants diagnosed postnatally, often by spontaneous delivery (76.9%). In the prenatal group 100% of newborns received prostaglandin E1 from day one, and the "postnatal" group at an average of 6th day. In the "prenatal" group the dose of PGE:  $0.02 \text{ ug / kg / min}$ , in the "postnatal" group the average dose was  $> 2 \times$  higher  $0,042 \text{ mg / kg / min}$ . A statistically significant relationship was shown: between the day of administering prostaglandin E1 and the number of postoperative recovery hospitalization days of the newborn with IAA.

Di George Syndrome occurred in 40% in the "prenatal" and in 15.4% in the "postnatal" group. Cardiac operations in the "prenatal" group were performed at average 19th day, and at average 21st day in the "postnatal" group. Infants in the "prenatal" group remained in the postoperative recovery room an average of  $7.85 + / -3.98$  days, and those from the "postnatal" group an average of  $8.38 + / -3.94$  days ( $p = 0.6212$ ). There were 3 deaths (7.3%): 1 patient diagnosed prenatally and 2 postnatally. Neonates with IAA in the "prenatal" group remained in the hospital on average:  $43.69 + / - 4.82$  days and  $39.54 + / -3.75$  days in the "postnatal" group. After cardiac surgery, hospitalization in the "prenatal" group amounted to an average of 25.2 days in the "postnatal" 26.7 days.

**Conclusions:** 1. The prenatal diagnosis of IAA was grounds to start the administration of prostaglandin E1 on the first day of life, and in the "postnatal" group the administration of prostaglandin followed on average 6 days after birth, the "postnatal" dose was  $2 \times$  higher. 2. A statistically significant correlation was demonstrated between the day of prostaglandin E1 commencement and the number of hospitalization days of the newborn with IAA in the postoperative recovery room, the length of hospital stay in the ICU and sometimes full hospitalization.

**Key words:** interrupted aortic arch, prenatal and postnatal diagnoses

## BACKGROUND

Interruption of the aortic arch (IAA - interrupted aortic arch) is the lack of continuity of the aorta in a portion of its arch<sup>1,2,3,4,5</sup>. It is a defect that requires systemic ductal dependent blood flow. The defect was first described in 1760 by M. Morgani<sup>1</sup>, for the first time subjected to surgi-

### How to Cite this Article:

Augustyniak A, Stodki M, Krajewski W, Moll J, Respondek-Liberska M. Comparison of the clinical status and the effects of treatment of newborns with interrupted aortic arch diagnosed prenatally and postnatally in the "Polish Mother's Memorial" Institute in Lodz in the years 2003-2012. *Prenat Cardio. 2014 Jun;4(2):11-19*



cal treatment in 1954<sup>3</sup> Interrupted aortic arch amounts up to 1.5% of all congenital heart defects in children<sup>1,2,3,4,7</sup>, according to other sources only 1% 5 75% of untreated newborns die in the first month of life, and the median survival time reaches 4 to 10 days<sup>1,3</sup>.

In the existing literature on prenatal cardiology there has not been an exhaustive comparative analysis

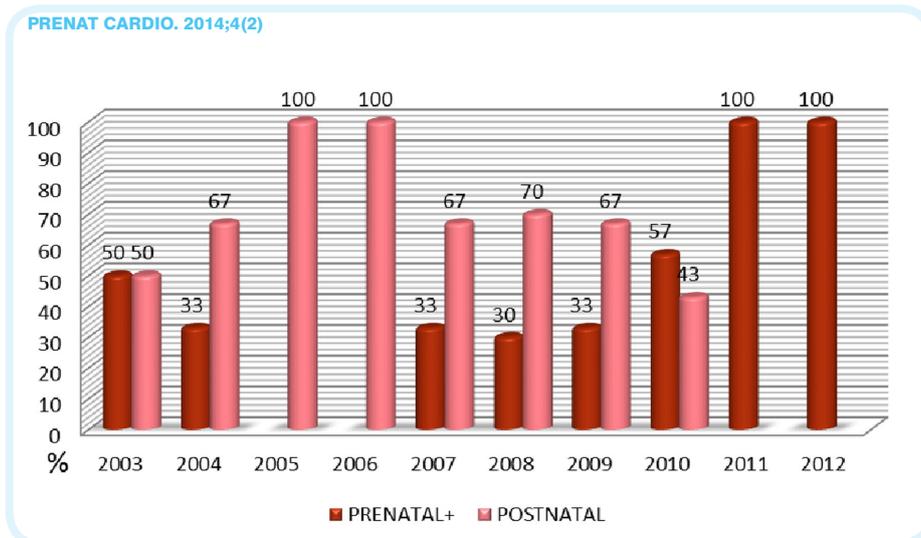


Figure 1. Comparison of neonates with IAA operated on in ICZMP through 2003-2012 period.

of the follow up of infants prenatally diagnosed with IAA and postnatally in the same medical facility.

**MATERIALS AND METHODS**

A retrospective analysis of medical records of 41 patients with congenital heart disease IAA undergoing treatment in the Department of Pediatric Cardiac Surgery, hospitalized in the Pediatric Department of Anesthesiology and Intensive Care ICZMP in Lodz in the years 2003-2012, including 15 after prenatal diagnosis in ICZMP.

We analyzed the following data: the size of the heart and cardiovascular endurance of the fetus, the mother's age during pregnancy, week of gestation at the time of delivery, mode of delivery, birth weight, number of points in the Apgar scale in the first minute of life, day of prostaglandin E1 commencement, day of life in which cardiac surgery was performed, extubation day after the operation, the number of days spent in the postoperative recovery room, the number of days in the Pediatric Intensive Care Unit (PICU), the length of hospitalization. Used for statistical analysis: Shapiro-Wilk test for the assessment of normal distribution and because in most groups the distribution of the measured traits was not consistent with normal distribution - for further analyzes nonparametric test was used: the Mann-Whitney test, Fisher's exact test and Pearson's correlation taking the level of significance to p-value = 0.05.

**RESULTS**

Annually, the Department of Cardiac Surgery in the years 2003-2012 ICZMP operated an average of four neonates with IAA: the most in 2008 - ten, the least in the year 2012 - one. In 2005 and 2006 100% of patients were newborns postnatally diagnosed with IAA. From 2010 the prenatal diagnosis of IAA was 57% of operated patients, and in 2011 and 2012 100% (Table 1, Figure 1).

In the past 10 years newborns with IAA after prenatal

diagnosis accounted for an average 36.6% of operated patients (Figure 2).

The defect in the form of IAA was diagnosed in 57% of mothers with high risk pregnancy - HRP (maternal age, poor obstetric history, family history). The average age of grvida with the diagnosis of fetal IAA was 29.25 + / -1.16 years (median 28, min 24 max 36)

The group, n = 14 fetuses with IAA heart size was within norm: an average of 0.33 + / -0.05 HA / CA (median 0.305, min 0.29 max 0.44).

Cardiovascular endurance in the 10 point Huhty CVPS scale (called Cardiovascular Profile Score) was 8 to 10 points (mean 9.56 + / -0.73, median 10, min 8 max 10).

In 10 fetuses with IAA (71.4%) non-cardiac anomalies were diagnosed: 6x polyhydramnios - (43%), 2x hypoplasia of the thymus (14.3%), and single other anomalies such as two-vessel umbilical cord (1x), IUGR (1x) and hyperechogenic multi-lobed placenta (1x). In the total group of neonates with IAA the majority (55.5%) was delivered vaginally. Infants diagnosed prenatally often were delivered by caesarean section (64.3%), and infants diagnosed postnatally more frequently were delivered vaginally (76.9%) (Figure 3). The difference was statistically significant (Fisher's exact test: p = 0.0377).

In the total group (n = 35) of infants with IAA the average gestational age at time of delivery was 38.86 + / -1.91 weeks (median 39, max 42 min 35), 33 infants (80.5%) were born on time, 2 at 37 weeks of gestation, 4 at 36 weeks and 2 at 35 weeks (a total of 19.5% of preterm births). In the group of infants with prenatal diagnosis 26.6% of deliveries took place preterm (U Mann-Whitney

YEAR	IAA	%	PRENATAL+	%	POSTNATAL	%
2003	2	5	1	50	1	50
2004	3	7,5	1	33	2	67
2005	2	5	-	-	2	100
2006	5	12,5	-	-	5	100
2007	6	15	2	33	4	67
2008	10	25	3	30	7	70
2009	3	7,5	1	33	2	67
2010	7	17,5	4	57	3	47
2011	2	5	2	100	-	-
2012	1	2,5	1	100	-	-
<b>SUMA</b>	<b>40</b>	<b>100</b>	<b>15</b>		<b>26</b>	

Table 1. Neonates with IAA operated in CZMP in 2003-2012 years

test:  $P = 0.2126$ ) (Figure 4).

The mean birth weight of newborns with IAA in the total group  $n = 39$  was 2924.36 g (median 3000, min 1450 max 3970) and no difference between the "prenatal" and postnatal " groups were found (Test Mann-Whitney  $U p = 0,7430$ ).

Apgar score according to the scale in the first minute after birth for all 39 neonates with IAA was  $8.2 \pm 1.19$  points (median 9, min 5 max 10) but in the "prenatal" group 8.13 points and in the "postnatal" group 9,08 points: the difference was statistically significant (U test Mann-Whitney test,  $p = 0.0106$ ) (Figure 5).

In all neonates with IAA treatment requiring persistent patency of the ductus arteriosus (constant intravenous infusion of prostaglandin E1) was administered: Infants in the "prenatal" group received prostaglandin E1 on the first day of life in 86% of cases, and in the "postnatal" group only 12% (Fisher's exact test  $p = 0.0000$ ) (Table 2, Figure 6).

The difference was also observed in average doses of prostaglandins: in the "prenatal" group of infants with IAA, it amounted to 0.02 ug / kg / min in the " postnatal" group average dose was more than twice as high 0,042 mg / kg / min.

Analysis based on the total hospitalization (number of days), length of stay in the postoperative recovery room and the time of hospitalization in the Pediatric Intensive Care Unit showed that there is a relationship between the day of administering the supply of prostaglandin E1 and length of hospitalization. A statistically significant correlation was shown: between the day of administering prostaglandin E1 and the number of hospitalization days the newborn with IAA spent in the postoperative recovery room, length of stay in the PICU and the complete hospitalization (Figure 7.8 and 9).

In the group of all newborns with IAA, 97.5% of infants had additional anomalies in the cardiovascular system: the most common ventricular septal defect (VSD), (92.7%) (Table 3).

For patients with IAA an additional anomaly (DiGeorge Syndrome) can also be present. In our series it appeared in 40% of cases in the prenatal group and in 15.4% in the "postnatal".

Neonates with IAA in the "postnatal" group were diagnosed in 69% of cases up to the eighth day of life. On the second day, 15.4% of newborns were diagnosed , on the third day 30.8% of newborns, after the age of eight days 30.6% of newborns were diagnosed (Figure 10). At the latest IAA was diagnosed at 21 days ( the additional malformations were TAC, VSD, ASD). On average, the diagnosis of IAA in the postnatal group was at 6,4 days. Invasive cardiac procedures among neonates with IAA were carried out in only two cases(4.9%) it was the Rashkind

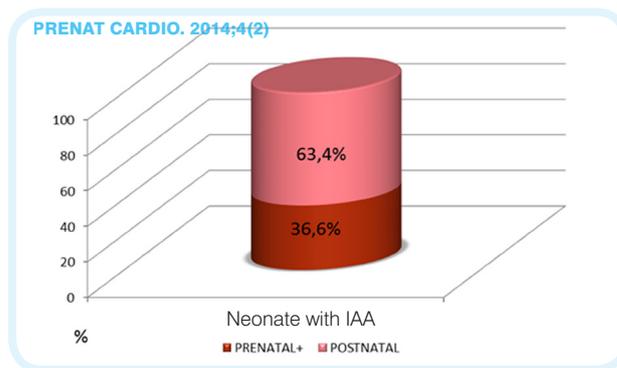


Figure 2. Percent of prenatal diagnosis of IAA

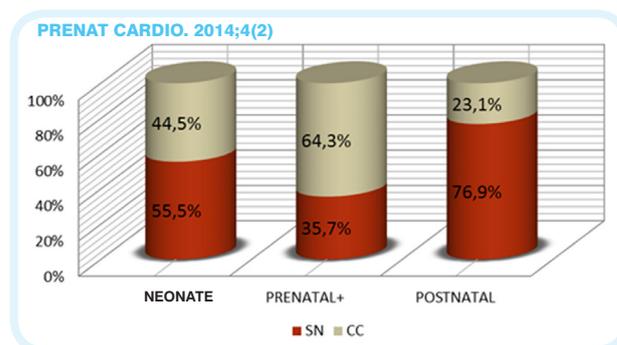


Figure 3. The way of delivery of neonates with IAA

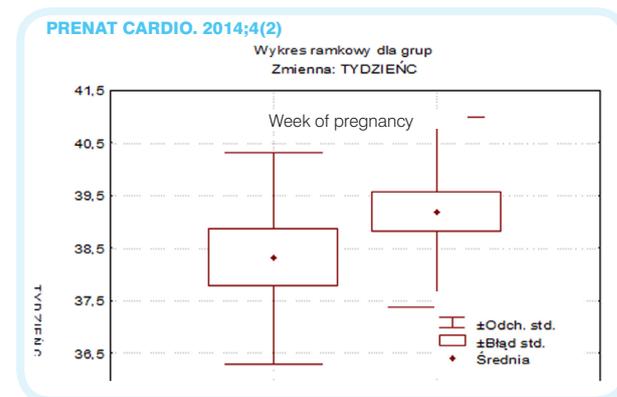


Figure 4. Gestational age at the delivery in pre- and postnatal group

Procedure, or balloon atrioseptostomy. Cardiac operations in 39 neonates with IAA were carried out on average  $20.82 \pm 9.56$  day (median 20, min 5 max 59). Neonates in the "prenatal" group were operated at an average age of 19.38 days, and postnatal group at the age of 21.4 days (Figure 11). The earliest surgical procedure was performed on day 5 in a newborn diagnosed postnatally. In this group was also the infant that underwent surgery the latest that is at 59 days (it was a premature baby of 36 weeks gestation and low birth weight 1450g, diagnosed at 11 days of life, treatment fatal). Among the group of "prenatal" IAA patients, cardiac surgery was performed the earliest at 10 days the latest at 35. The correlation between the time of the operation and the number of days

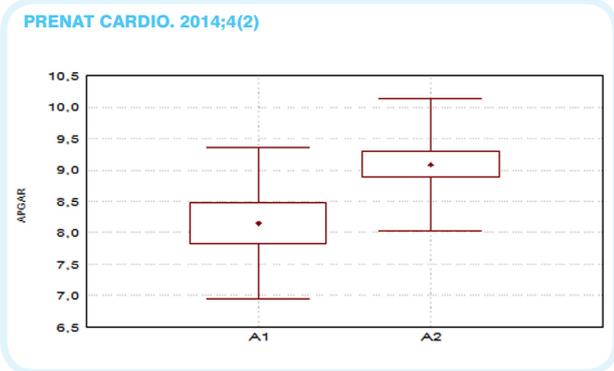


Figure 5. Comparison of Apgar score in newborns with IAA in pre- and post-natal diagnosis

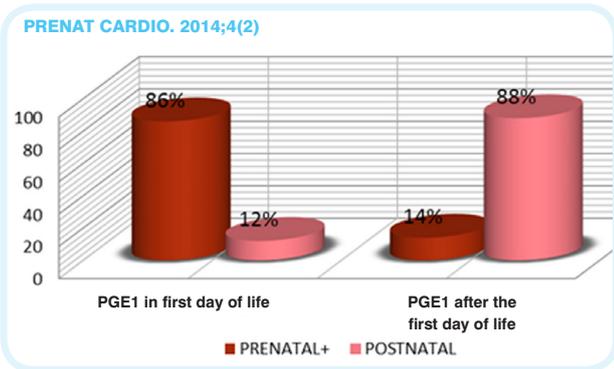


Figure 6. The day of implementation of prostaglandin E1 in pre- and postnatal group

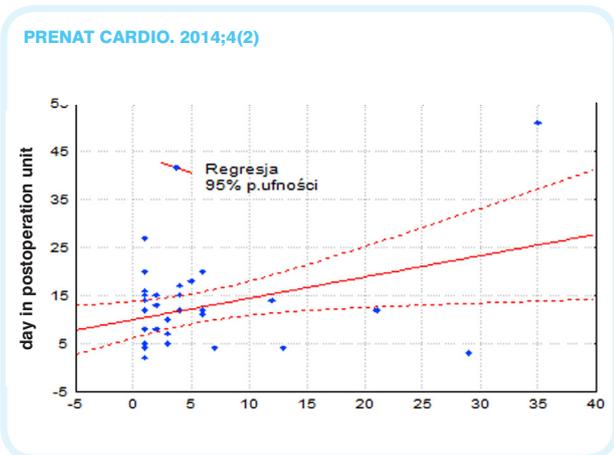


Figure 7. Correlation between postoperation unit stay and first day of prostaglandin E1 implementation in newborn with IAA

of hospitalization of newborns with IAA was demonstrated (Pearson correlation,  $p < 0.05$ ) (Figure 12).

Neonates ( $n = 13$ ) with IAA in the “prenatal” group remained in the postoperative recovery room an average of  $7.85 \pm 3.98$  days (median 7, min 4 max 16), and in the “postnatal” group  $n = 26$ , mean  $8.38 \pm 3.94$  days (median 7, min 3 max 18), (U Mann-Whitney test,  $p = 0.6212$ ). Over the analyzed 10 years neonatal deaths were recorded in three cases of IAA (7.3%): 1 patient diagnosed prenatally (6.6%), and 2 postnatally (7.7%). Duration of hospitalization was analyzed in three periods: the total stay beginning with admission to hospital to discharge

PROSTAGLANDIN E1 IMPLEMENTATION DAY	PRENATAL +	%	POSTNATAL	%
1	13	86	3	12
2-3	1	7	7	26
4-8	1	7	9	35
9-14	-		6	23
>14	-		1	4

Table 2. Day of prostaglandin E1 implementation in newborn with IAA in pre- and postnatal diagnosis.

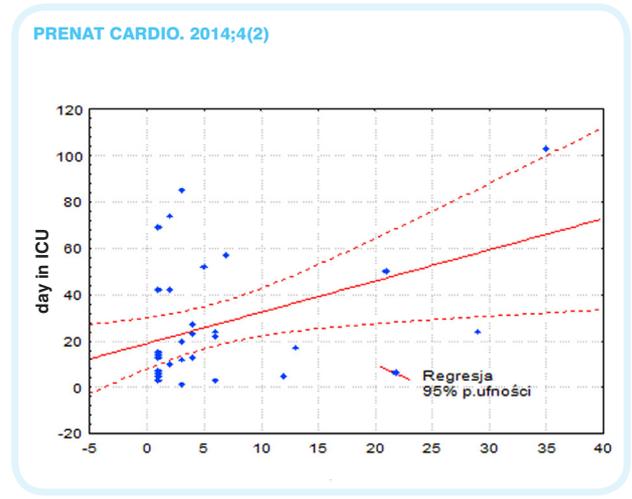


Figure 8. Correlation between stay at the intensive care unit and the first day of implementation of PGE1 in group of newborns with IAA

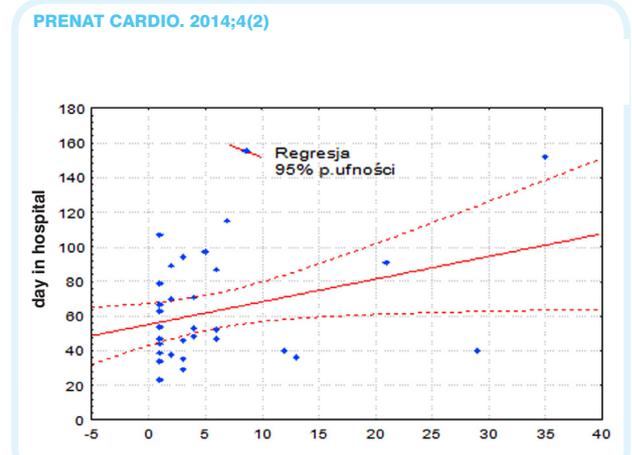


Figure 9. Correlation between days of hospital stay and first day of implementation of PGE in newborns with IAA

from ICZMP, from the date of surgery to discharge and from admission to day of operation. Infants diagnosed prenatally had a longer time of hospitalization, they awaited longer for cardiac surgery, while the period from surgery to discharge was shorter. Neonates with IAA remained in the hospital on average:  $43.69 \pm 4.82$  days, prenatal group + “( $n = 13$ , median 37, min 24 max 79) and  $39.54 \pm 3.75$  days postnatal group, “( $n = 26$ , median 34.5, min 16 max 91.), (Figure 13). We compared the total duration of hospitalization of newborns with IAA with prenatal diagnosis and without: “prenatal” Group awaited

COEXISTENT CARDIAC ANOMALIES	NEONATE WITH IAA	%
VSD - Ventricular septal defect	38	92,7
ASD - Atrial septal defect	15	36,5
LVOTO – Left ventricular outflow tract obstruction	5	12,2
TGA – Transposition of the great artery	3	7,3
CAV – Atrioventricular canal	1	2,4
DORV – Dual outflow right ventricle	2	4,8
FOA – Persistent foramen ovale	5	12,2
HMV –Hypoplastic mitral valve	2	4,8
HLV – Hypoplastic left ventricle	3	7,3
HRV – Hypoplastic right ventricle	1	2,4
HAA – Hypoplastic aortic arcg	1	2,4
HAoV –Hypoplastic aortic valve	5	12,2
AAoV – Aortic valve atresia	1	2,4
Taussig-Bing syndrome	1	2,4
Truncus arteriosus communis	1	2,4

Table 3. Coexisted cardiac anomalies with IAA in newborns operated on in ICZMP in 2003-2012

for treatment on average 18.5 days, "postnatal" group an average of 12 days. After cardiac surgery, hospitalization in the "prenatal" group amounted to an average of 25.2 days in the "postnatal" 26.7 days (Table 4, Figure 14).

## DISCUSSION

Interruption of the aortic arch is probably formed as a result of two mechanisms: disorders of cardiac neural crest cell migration and hemodynamic disorders<sup>8</sup>. Neural crest cells contribute to the formation of the arteries of aortic arches. Their impaired migration to the cardiac neural tube has a significant impact on the process. Abnormal development of the sinistral fourth aortic arch causes the left subclavian artery not to reach the desired position (the formation of IAA Type B)<sup>3</sup>. Hemodynamic disorders are the result of reduced blood flow through the aortic arch, which during embryogenesis directly influences the evolution and the expansion of vessels<sup>3,8,9</sup>. Currently, thanks to prenatal ultrasound testing obstetricians can detect abnormal images of the mediastinum, and in the prenatal cardiology referral center make a detailed analysis of heart defects<sup>9</sup>. The most important marker of interrupted aortic arch is the broadening of the pulmonary artery in the mediastinum<sup>9,10</sup>, which is a result of increased blood flow through the vessel. Measuring the width of the section of the pulmonary artery PA and aorta Ao and calculating the index PA / Ao (standard 1.09, 1.18 +/- 0.06) allow the confirmation of the correct relations between the diameters of the large vessels or to detect heart defects<sup>9,11,12,13</sup> (Figure 1). PA / Ao ratio may be helpful in differentiating IAA with aortic isthmus stenosis or aortic arch hypoplasia<sup>10,12</sup>. The values of PA / Ao > 3 may indicate the place in

which the aorta is interrupted, indicating type B<sup>11,12</sup>. The specific image for interrupted aortic arch is a straight, narrow ascending aorta cephalad and visualization of the "Y" symptom<sup>9,12,14</sup>. Differences in the image acquired are dependent on the type of IAA defect: type B is the straight, narrow aorta, other types of defects in their image show a slight curvature associated with the continuity of the aortic arch segment<sup>12</sup>. The use of new imaging techniques (multi-level 3D and 4D imaging, spatial-temporal correlation image SITC) contribute to the improvement of results gained by prenatal diagnosis and help in differentiating the type of defect<sup>12,15,16</sup>. The analysis of the mediastinum also allows us to evaluate the thymus, as the aplasia or hypoplasia may suggest chromosomal abnormalities (microdeletion 22q11)<sup>10,17</sup>. Imaging of the aortic arch is relatively difficult technically, described simply as a challenge to echocardiologists<sup>9</sup>. Because the detection and diagnosis of IAA in the fetus is difficult<sup>14</sup>, the majority of newborns with this congenital heart malformation are born without prenatal diagnosis, not manifesting signs of this defect in the first days of life. Pediatric cardiologists (including Govindaswami et al 2012, Altman et al 2013) assume that a critical heart defect is one that requires the intervention of cardiac surgery during the first year of life<sup>18,19</sup>. Other authors like Szymkiewicz-Dangel 2008 and Ewer et al 2012 define critical heart defects as those that require intervention within a month of birth<sup>6,20</sup>. In contrast, prenatal cardiologists that created the division stated by ORPKP 2012 identify critical and severe heart defects among newborns. In the case of critical defects the newborn should be transported from the delivery room straight to the catheter lab or operating room because intervention is necessary in the first hours of life. However, in the case of severe defects interventional treatment or surgical closure can be performed electively in the newborn<sup>21,22,23,24</sup>. Analysing the follow up of neonates with

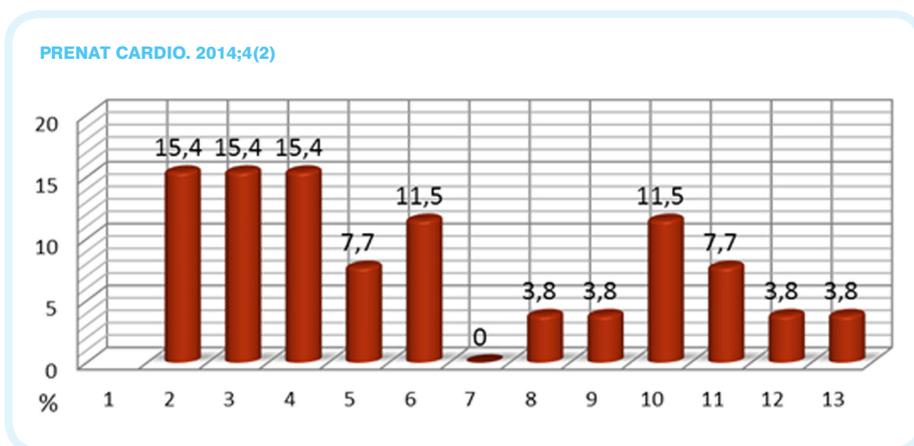


Figure 10. Days of postnatal diagnoses of IAA

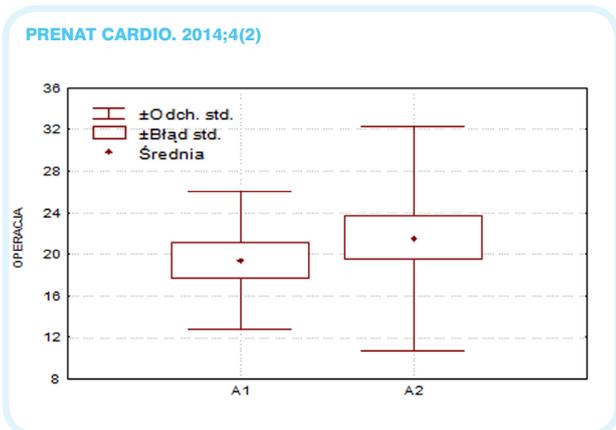


Figure 11. The day of cardiac surgery in neonates with IAA in two groups of newborns: with prenatal and postnatal diagnoses

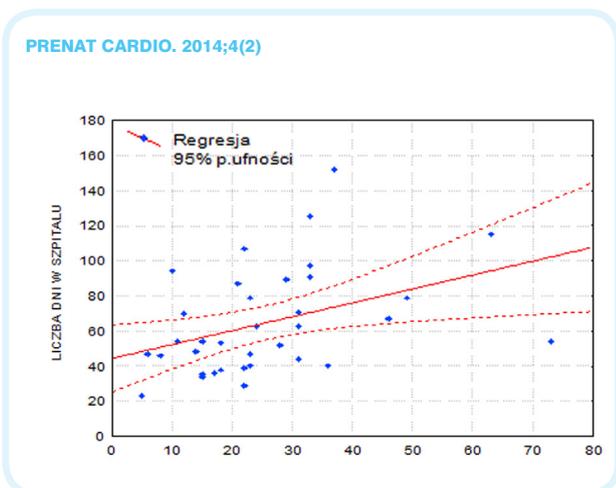


Figure 12. Correlation between days of hospital stay and day of cardiac surgery in newborns with IAA

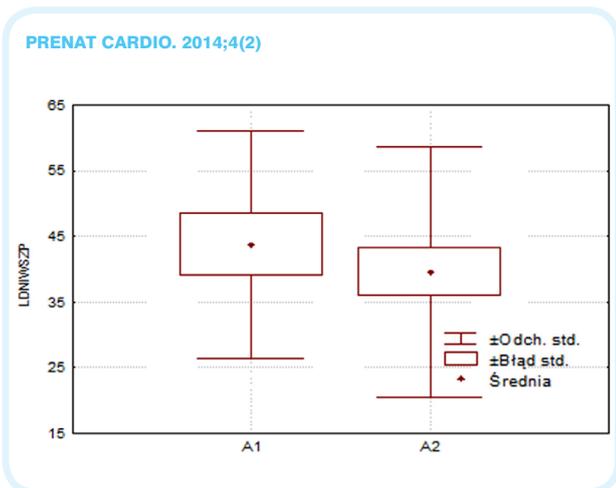


Figure 13. Days of hospital stay in neonates with IAA in group of prenatal diagnoses and in group of postnatal diagnoses

interrupted aortic arch that underwent surgery at the Institute of Polish Mother’s Memorial Hospital in Lodz in the years 2003-2012, we observed that these procedures were carried out between 5 to 28 days of age, and therefore

the IAA in most cases classified as severe heart defect (not critical). Prenatal detection of the defect (IAA) at the ZDiPWW over the years from 2003 to 2012 significantly increased an average of 36.6%, as well as in other centers: Boston 24%: increase in detection from 11% to 43% in the period 1988-2009 (Vogel et al 2010)<sup>25</sup>; In Giesen in the years 1994-2010 28.6% of newborns with IAA were diagnosed prenatally (Axt-Fliedner et al 2011)<sup>14</sup>. Our result: better than the foreign data probably due to a later period of analysis, better technical capabilities walking hand in hand with the increasing experience of doctors. In our series of cases high-risk pregnancies predominated (57%), probably because it is the highest referral center for fetal anomalies from all over the country. Fetuses with IAA had normal heart size, displayed good intrauterine cardiovascular efficiency, (8 - 10 points scale Huhty - CVPS). Wieczorek et al 2008 study from Florida showed that CVPS score  $\geq 8$  indicates that the infant will be significantly less exposed to perinatal mortality than the result of  $<7$ <sup>26</sup>. Neonates from the analyzed data prenatally did not present any signs indicating perinatal threat, and so from the standpoint of prenatal cardiology these defects were not critical. In the analyzed group of all neonates 45.5% of births required intervention, but in the group that underwent prenatal diagnosis, 64.3% of births took place by caesarean section. This suggests either excessive preventative measures taken by obstetricians or pressure from the gravida and probably the present knowledge of a fetal anomaly that were cause against taking the risk of natural delivery. According to the Lodz School of prenatal cardiology newborns with severe heart defects do not have to be delivered invasively (which is desirable and recommended for neonates with prenatally diagnosed critical defects)<sup>21,22,23,24</sup>. Average Apgar score in the first minute of life for the entire group of neonates with IAA: 8.2 points confirms that the fetal circulation effectively protects against the deterioration of the general condition immediately after birth. Newborns born prematurely with low birth weight received less points. In this analysis we demonstrated a statistically significant difference in lower Apgar score for neonates with prenatally diagnosed IAA compared to newborns with IAA diagnosed postnatally ( $p = 0.0106$ ). This result could be affected by the fact that infants prenatally diagnosed with IAA were born earlier (26.6% preterm births) and with lower birth weight (2820g vs median. 3025g) than infants in the “postnatal” group. The mean birth weight of newborns with IAA was 2924 g (median 3000+/-504.76;1450-3970). In the data of other authors weight of infants during surgery was stated: 3000g in Chicago (1,7-6,1 kg) by Morales et al 2011, also on average 3000g (1.8-20kg), France (Serraf et al 1996)<sup>27,28</sup>. All newborns with IAA before cardiac surgery received an infusion of prostaglandin E1 to maintain the patency of the ductus arteriosus. In the groups of infants “prenatal+” and “postnatal” differences concerned the day of commencement of the drug: in the prenatal group 86% of infusions were administered on the first day (100% up to day eight), and in the “postnatal” group in only 12%

of cases (73% to the eighth day, the others later). Many authors emphasized the need for administration of prostaglandins in neonates with IAA (Morales et al 2006 Serraf et al 1996, Jacobs et al 1995)<sup>27,28,29</sup>, but cited the work does not include the dose of PGE1 or was the time of commencement of the drug analyzed. In our material for the first time we showed that neonates from the "prenatal" group received an average of about half the dose of prostaglandin E1 compared to neonates from the "postnatal" group (IAA "prenatal +" 0.02 g / kg / min vs "postnatal" 0,042 mg / kg / min), which provided a lower risk of side effects of treatment. It was also shown that the earlier administration of PGE1 decided a shorter stay of newborns with IAA in the postoperative recovery room, the PICU and a shorter hospital stay in general (Pearson correlation,  $p < 0.05$ ). At the Institute CZMP newborns with IAA underwent surgery approximately at 20.82 days (from 5 to 59 days), which puts our center far behind other institutions. In France, at the Marie-Lannelongue Hospital data derived from a ten year observation suggest that newborns with IAA underwent surgery at an average of 9 days of life (from day 1 to 6 years) (Serraf et al 1996)<sup>28</sup>, in a hospital in Ohio, the median operation was at 8 days (2 to 271 days) (Morales et al 2006)<sup>27</sup>, Canada: 7 days (1 to 60 days) (McCrindle et al 2005)<sup>30</sup>, but Giessen the average day of operation was 17.1 days (from 1 to 210 days)<sup>14</sup>. The largest ever conducted analysis of the follow up of newborns with IAA operated in Canada showed that the older the age at the time of surgical correction of the defect poses a significant higher risk of intervention in the future (McCrindle et al 2005)<sup>14</sup>. It was a study covering 16 years of the performed surgery. In the presented material we analyzed the factors in the periprocedural period. Examined the correlation between time of hospitalization: in the recovery room, at the PICU in the hospital on general - from the day in which cardiac surgery was performed. A statistically significant ( $p < 0.05$ ) correlation is presented: the earlier the operation is performed the shorter the length of hospital stay (measured in days of hospitalization). It has been shown that infants diagnosed "postnatally" were operated earlier than those prenatally (19.38 vs 21.4 days, insignificant difference). Surprisingly, despite the early diagnosis newborns had to wait for the correction of defects a dozen or even more than twenty days, at a lead pediatric cardiac surgery center in Poland. Our center collects a very large group

	TOTAL HOSPITALIZATION	POSTOPERATIVE HOSPITALIZATION	PREOPERATIVE HOSPITALIZATION
PRENATAL +.	43,7	25,2	18,5
POSTNATAL	39,5	26,7	12,8

Table 4. Average total hospitalization, pre- and postoperative.

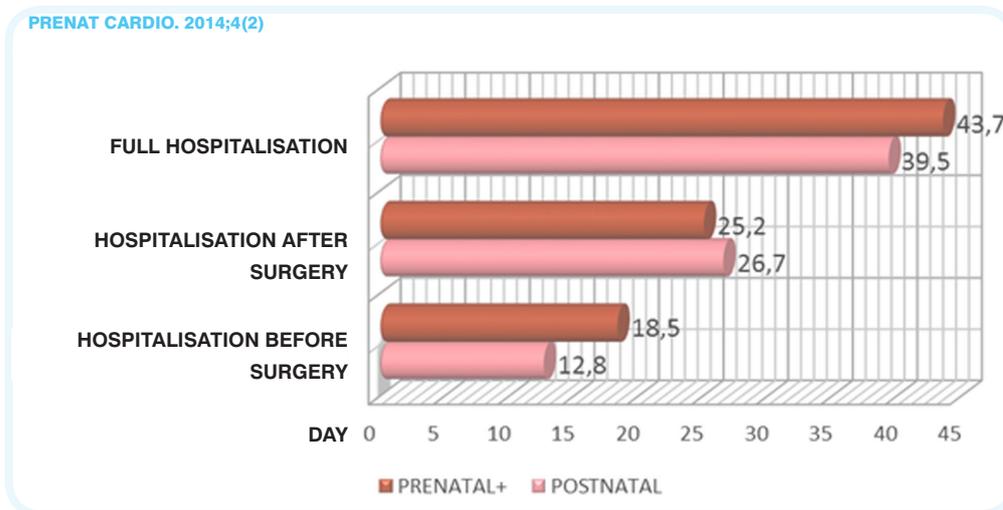


Figure 14. Days of hospital stay in prenatal and postnatal groups: total stay, stay after surgery, stay before surgery

of patients, and thus creates a waiting list for cardiac surgery. Newborns provided with prostaglandin E1 treatment often "await" for the correction of defects, giving way to neonates diagnosed postnatally that are in worse clinical condition. The analyzed material shows that neonates with IAA in the "prenatal" group remained in the postoperative recovery room shorter (an average of 7.85 days) than infants in the "postnatal" (8.38 days). But the difference was insignificant ( $p = 0.6212$ ). Postoperative mortality of newborns with IAA at the ICZMP center was low 7.3% (3 deaths, including 1 death of a newborn prenatally diagnosed), according to Serraf et al 1996 18.9%<sup>28</sup>, Axt-Fliedner et al, 2011:<sup>18</sup> 14%, and 7% by Morales et al 2006 27. Already cited the largest analysis in Canada, on 472 neonates with IAA presents a sixteen year observation of the follow up of the operated children: 59% of patients achieved an overall survival of 16 years after surgery. (McCrindle et al 2005)<sup>30</sup>. According to data from the literature risk factors for death in neonates with IAA were: low weight and premature birth, old age of the neonate upon surgery, many congenital anomalies, DiGeorge syndrome, a concomitant LVOTO (Morales et al 2006 McCrindle et al 2005, Mr. et al 2005)<sup>27,30, 31</sup>. Time of hospital of newborns with IAA in the analyzed material from ICZMP was an average of 40.9 days. It was longer than that of a newborn with IAA in the hospital in Hamburg, where the average was 25,5-27,3 days (Lacour-Gayet et al 2004)<sup>32</sup> It is interesting that infants prenatally diagnosed with IAA remained in the hospital longer than those diagnosed postnatally (43.69 vs. 39.54 days). Length of hospital stay was divided into two periods: given the time from admission to surgery and after surgery up until

discharge from hospital. It has been shown that the "prenatal" group had a slightly shorter hospital stay after cardiac surgery (25.2 vs. 26.7 days), Length of hospitalization of newborns in the "prenatal +" group was prolonged due to a longer stay in the hospital before surgery (18.5 vs. 12 days). The presented material demonstrates DiGeorge Syndrome in 24.4% of neonates with IAA operated in ICZMP. Among those operated in Ohio, DiGeorge syndrome was diagnosed in 35% of newborns with IAA (Morales et al 2006). The presence of 22q11 microdeletion is an important risk factor for mortality<sup>27</sup>. Axt-Fliedner et al 2011 showed the presence of DiGeorge Syndrome only in the group of neonates with IAA type B in 75%<sup>14</sup>. In our center far more chromosomal abnormalities type 22q11 microdeletion were observed in newborns prenatally diagnosed (40% vs. 15.4%). Perhaps this is an additional key to explain why newborns prenatally diagnosed had a similar postnatal course (despite early administration of prostin) in relation to the "postnatal, with late administration of prostin.

## CONCLUSIONS

Prenatal diagnosis of IAA was grounds for early administration of prostaglandin E1 on the first day of life, in the "postnatal" group commencement of prostaglandin followed on average 6 days after delivery (statistically significant difference Fisher's exact test:  $p = 0.0000$ ), while in the "postnatal" group the dose was 2 x higher.

A statistically significant correlation between the day of prostaglandin E1 commencement and the number of days of hospitalization of the newborn with IAA in the postoperative recovery room, the length of hospital stay in the PICU and the overall time of hospitalization.

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#### **Conflict of interest**

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