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RADIOGRAPHIC SEMIOTICS OF CONGENITAL HEART DEFECTS COMPLICATED BY HIGH PULMONARY HYPERTENSION: DIAGNOSTIC CRITERIA AND CLINICAL SIGNIFICANCE

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Abstract

Relevance of the study:

Congenital heart defects (CHD) occupy one of the leading places among congenital anomalies of development and are a significant cause of infant mortality and disability in the child population worldwide: according to the World Health Organization, the incidence of CHD is 8–12 cases per 1000 live births, and about 1.35 million children with this pathology are born annually in the world [1, 2]. Among all forms of CHD, defects are of particular clinical importance, accompanied by blood discharge from left to right, such as ventricular septal defect, atrial septal defect, patent ductus arteriosus, and atrioventricular canal, which together account for up to 40–50% of all CHD and, in the absence of timely surgical correction, are naturally complicated by the development of pulmonary arterial hypertension (PAH) [3, 4]. According to large epidemiological studies, CHD-associated PAH is detected in 1.6–12.5 per million population, while in patients with uncorrected malformations with pulmonary hypervolemia, pulmonary hypertension is formed in 30–50% of cases within the first two years of life [5, 6]. An extreme and irreversible manifestation of this complication is Eisenmenger syndrome, in which the pressure in the pulmonary artery reaches the systemic level, blood discharge becomes right-left and surgical correction of the defect is impossible; The survival rate of such patients is significantly reduced, and the median life expectancy does not exceed 40–50 years [7, 8]. In

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the context of the widespread introduction of echocardiography, multispiral computed tomography and catheterization of the heart cavities, chest X-ray is often underestimated as a diagnostic tool, but it remains the primary, generally available and highly informative method, which allows, based on the analysis of the pulmonary vascular pattern, the size and configuration of the shadow of the heart, the state of the pulmonary trunk and the nature of changes in the pulmonary parenchyma, to suspect the presence of CHD, to assess the degree of hemodynamic disorders and stratify patients according to the urgency of further examination and surgical treatment [9, 10]. At the same time, systematized data on the radiographic semiotics of CHD complicated by high pulmonary hypertension are not fully presented in domestic and foreign literature, and diagnostic criteria based on quantitative and qualitative radiological signs require clarification and standardization taking into account modern clinical guidelines [11, 12, 13, 14].

The aim of this study is to systematize and analyze the radiographic signs of congenital heart defects complicated by high pulmonary hypertension, to determine their diagnostic significance and to develop a structured algorithm for X-ray assessment to optimize the clinical management of this category of patients.

Materials and methods:
Chest X-ray (CRX) was a mandatory component of the diagnostic algorithm in all 123 patients included in this study. Imaging was performed in standard projections — direct (anteroposterior) and left lateral — under conditions of maximum possible inspiration, taking into account age and clinical limitations. In infants of the neonatal period and early infants (n=41; 33.3%), radiographs were obtained in the prone position at the back due to the severity of the condition; in patients over 6 months of age (n=82; 66.7%) — mainly in an upright position.

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Results:

The cardiothoracic index was calculated as the ratio of the maximum transverse diameter of the cardiac shadow to the maximum internal diameter of the chest at the level of the right dome of the diaphragm. Normally, in children under 3 years of age, the CTI does not exceed 0.55. The measurement results depending on the nosological form are presented in Table 1.

Table 1. Cardiothoracic index values depending on the nosological form of CHD

Nosological form	n	CTI (M±SD)	Range	CTI ≥0.60, n (%)
VSD insulated	99	0,64±0,07	0,52–0,81	78 (78,8)
VSD + CoAo	2	0,67±0,04	0,64–0,70	2 (100,0)
VSD + Pr. Ao	2	0,63±0,06	0,59–0,67	1 (50,0)
DOMS from RV	7	0,68±0,05	0,61–0,75	7 (100,0)
TADLV	6	0,61±0,08	0,52–0,72	4 (66,7)
AVK (Full Form)	4	0,71±0,06	0,65–0,78	4 (100,0)
TMS	1	0,73	—	1 (100,0)
DOMS+AVK+TADLV	1	0,76	—	1 (100,0)
Taussig-Bing anomaly	1	0,69	—	1 (100,0)
Total	123	0,65±0,07	0,52–0,81	99 (80,5)

Note: M is the average; SD is the standard deviation; CoAo — coarctation of the aorta; Pr. Ao — right aorta; DOMS — double discharge of the great vessels; TADLV – total abnormal pulmonary vein drainage; AVK — atrioventricular canal; TMS — transposition of the great vessels.

Cardiomegaly (CTI ≥0.60) was recorded in 99 of 123 patients (80.5%). The highest values of CTI were observed in the complete form of VKA (0.71±0.06), TMS (0.73) and combined malformation DOMS + VKA + TADLV (0.76), which reflects a pronounced volumetric overload of all heart chambers in these nosologies. In isolated VSD, the mean CTE was 0.64±0.07, and in 78 of 99 patients (78.8%) of this group, the CTI exceeded the threshold value of 0.60.

Statistically significant differences in CTI between groups with different nosological forms were confirmed by univariate analysis of variance (ANOVA): F(8.114)=3.42; p=0,0014. Pairwise comparison by the Tukey method revealed

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significant differences between the group of isolated VSD and the VKA group (p=0.031), as well as between VSD and TMS (p=0.048).

Analysis of the configuration of the cardiac shadow made it possible to identify several characteristic patterns presented in Table 2.

Table 2. Configuration of the cardiac shadow in various nosological forms of CHD with LGV

Configuration	Characteristics	Prevailing nosologies	n (%)
Mitral	Waist smoothing, bulging of the second arch on the left, displacement of the top to the left and down	VSD, AVK	71 (57,7)
Globular	Uniform magnification of all chambers, rounded shadow	AVK, DOMS+AVK+TADLV	18 (14,6)
"Egg on the side"	Narrow vascular bundle, oval shadow	TMS	1 (0,8)
"Snowman" (figure "8")	Dilation of the upper mediastinum due to an abnormal venous collector	TADLV (supracardial form)	3 (2,4)
Mixed/atypical	Combining features of multiple configurations	DOMS from PJ, Taussig-Bing	30 (24,4)
Total			123 (100,0)

The mitral configuration of the cardiac shadow was the most common (57.7%) and was mainly due to dilatation of the left atrium and left ventricle during left-right blood bypass grafting. Bulging of the second arch along the left contour of the heart, corresponding to the dilated trunk of the pulmonary artery, was recorded in 89 patients (72.4%), which is an indirect radiographic marker of pulmonary hypertension.

The state of the pulmonary vascular pattern was assessed taking into account standardized criteria. An increase in the pulmonary pattern was regarded as pathological when vascular shadows were visualized in the outer third of the pulmonary fields, where they are not normally detected. Dilation of the lung roots was assessed by the diameter of the right descending pulmonary artery (PNLA):

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a value of more than 1.2 tracheal diameter in children under 1 year of age and more than 1.0 in children 1–3 years of age was considered pathological.
Quantitative characteristics of the radiographic signs of LGV are presented in Table 3.

Table 3. Frequency of radiographic signs of pulmonary hypertension in the examined patients

Radiographic sign	Abs. Number	%	95% CI
Enhancement of the pulmonary vascular pattern	118	95,9	90,6–98,6
Expansion of lung roots	112	91,1	84,6–95,4
Bulging of the pulmonary artery arch (II arch on the left)	89	72,4	63,5–80,1
Cardiomegaly (CTI ≥0.60)	99	80,5	72,3–87,1
Signs of venous stasis	47	38,2	29,6–47,4
Interstitial pulmonary edema	31	25,2	17,9–33,9
Alveolar pulmonary edema	9	7,3	3,4–13,4
Right-sided position of the aortic arch	8	6,5	2,8–12,4
Sign of a "snowman"	3	2,4	0,5–6,9
Hydrothorax (unilateral or bilateral)	14	11,4	6,4–18,4

Note: 95% CI is a 95% Wilson confidence interval.
An increase in the pulmonary vascular pattern was detected in the overwhelming majority of patients (118 out of 123 (95.9%). In 5 patients with obstructive TADLV, the pulmonary pattern was normal or moderately impoverished, which is due to obstruction of venous outflow and redistribution of blood flow. Dilation of the lung roots was recorded in 112 patients (91.1%), while in 78 of them (69.6%) the dilation was bilateral symmetrical, in 34 (30.4%) it was asymmetrical, with a predominance of right-sided dilation.

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Signs of venous stasis (redistribution of blood flow to the upper lobes, dilation of the pulmonary veins, Curley lines type B) were found in 47 patients (38.2%). This sign was significantly more common in TADLV (5/6; 83.3%) and VKA (3/4; 75.0%) compared to isolated VSD (34/99; 34.3%; χ^2=8.74; p=0.003 and χ^2=6.12; p=0.013, respectively).

For an in-depth analysis of the radiographic correlates of LGV, the patients were stratified by the degree of pulmonary hypertension according to cardiac catheterization and echocardiography: moderate LGV (mean pulmonary artery pressure – Mean 25–45 mm Hg; n=38), high LGD (Mean LDLA 46–70 mm Hg; n=61), and ultra-high LGD (Mean LDLA >70 mm Hg; n=24).

Table 3.2.4. Radiographic indicators depending on the degree of pulmonary hypertension

Indicator	Moderate LGV (n=38)	High LGV (n=61)	Ultra-high LGV (n=24)	p
CTI (M±SD)	0,61±0,05	0,65±0,07	0,69±0,08	0,001*
Amplification of the l/s of the pattern, n (%)	38 (100,0)	61 (100,0)	19 (79,2)	0,001**
Root expansion, n (%)	32 (84,2)	57 (93,4)	23 (95,8)	0,187**
Arch II bulge, n (%)	21 (55,3)	47 (77,0)	21 (87,5)	0,009**
Venous congestion, n (%)	8 (21,1)	22 (36,1)	17 (70,8)	<0.001**
Interstitial edema, n (%)	3 (7,9)	16 (26,2)	12 (50,0)	<0.001**
Alveolar edema, n (%)	0 (0,0)	4 (6,6)	5 (20,8)	0,003**
PNLA/Tracheal Diameter (M±SD)	1,18±0,14	1,41±0,18	1,67±0,22	<0.001*

*Note: * is the Kruskel-Wallis criterion; ** is the χ^2 criterion or Fisher's exact criterion; l/s — pulmonary-vascular; PNLA is the right descending pulmonary artery.*

A significant direct correlation was established between the degree of LGH and the value of the CTE (r=0.47; p<0.001), as well as between the degree of LGV and the ratio of the diameter of the PNLA to the diameter of the trachea (r=0.68; p<0.001). The latter indicator demonstrated the highest diagnostic value among

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all the evaluated radiographic parameters: at the threshold value of ≥1.50, the sensitivity in detecting ultra-high LGH was 79.2%, specificity was 84.8%, and the area under the ROC curve (AUC) was 0.86 (95% CI: 0.76–0.93; p<0.001).

It is noteworthy that in ultra-high LGV, the increase in the pulmonary pattern was significantly less common (79.2%) than in moderate and high LGV (100.0% in both groups; p=0.001). This phenomenon is explained by the development of obstructive pulmonary vasculopathy with a reduction in peripheral pulmonary blood flow, which is an unfavorable prognostic sign and indicates irreversible changes in the pulmonary vascular bed.

VSD with high pulmonary hypertension (n=99). The radiographic picture was characterized by a mitral configuration of the cardiac shadow (n=71; 71.7%), a pronounced increase in the pulmonary vascular pattern (n=97; 98.0%), dilation of the lung roots (n=91; 91.9%), and cardiomegaly (n=78; 78.8%). Bulging of the second arch along the left contour, corresponding to the dilated trunk of the pulmonary artery, was recorded in 68 patients (68.7%). In 12 patients (12.1%) with VSD and concomitant CoAo or right aortic arch, additional signs were revealed: expansion of the shadow of the upper mediastinum, asymmetry of the pulmonary pattern.

DOMS from the right ventricle (n=7). The X-ray picture was distinguished by significant variability due to the anatomical features of the relationship between the great vessels. Cardiomegaly was detected in all 7 patients (100%), the CTI averaged 0.68±0.05. In 5 patients (71.4%), a mixed configuration of the cardiac shadow with signs of enlargement of the right parts was determined. An increase in the pulmonary pattern was recorded in 7 patients (100%), protrusion of the second arch in 6 patients (85.7%).

TADLV (n=6). Radiographic features differed significantly depending on the anatomical shape. In the supracardial form (n=3), a pathognomonic picture of the "snowman" or "figure 8" was revealed, due to the dilation of the vertical vein and the left ring vein. In the infracardial form (n=2), signs of pronounced venous stasis with interstitial pulmonary edema and impoverishment of the pulmonary pattern were determined. In the mixed form (n=1), the radiographic picture was atypical. The mean CTI value for TADLV was 0.61±0.08.

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VKA complete form (n=4). The most pronounced cardiomegaly in the studied cohort: CTI 0.71±0.06. Spherical or mitral configuration of the heart shadow with signs of enlargement of all four chambers. Pulmonary pattern enhancement and expansion of lung roots were observed in all 4 patients (100%). Signs of venous stasis were observed in 3 patients (75.0%).

TMS (n=1). The classic picture of the "egg on the side" with a narrow vascular bundle in the anteroposterior projection, due to the parallel location of the aorta and pulmonary artery. CTI 0.73. A pronounced increase in the pulmonary pattern.

Taussig-Bing anomaly (n=1). Radiographically pronounced cardiomegaly (CTI 0.69), increased pulmonary pattern, expansion of the lung roots, signs of enlargement of the right parts of the heart.

Summary data on nosolological-specific radiographic signs are presented in Table 3.2.5.

Table 3.2.5. Nosoa-specific radiographic signs of CHD with LGV

Sign	VSD (n=99)	DOMS for RV (n=7)	TADLV (n=6)	AVK (n=4)	Other (n=7)
Cardiomegaly (CTI≥0.60), n (%)	78 (78,8)	7 (100,0)	4 (66,7)	4 (100,0)	6 (85,7)
Mitral configuration, n (%)	71 (71,7)	2 (28,6)	1 (16,7)	2 (50,0)	1 (14,3)
Spherical configuration, n (%)	9 (9,1)	2 (28,6)	0 (0,0)	2 (50,0)	5 (71,4)
Amplification of the l/s of the pattern, n (%)	97 (98,0)	7 (100,0)	4 (66,7)	4 (100,0)	6 (85,7)
Root expansion, n (%)	91 (91,9)	7 (100,0)	5 (83,3)	4 (100,0)	5 (71,4)
Arch II bulge, n (%)	68 (68,7)	6 (85,7)	3 (50,0)	4 (100,0)	8 (100,0)
Venous congestion, n (%)	34 (34,3)	2 (28,6)	5 (83,3)	3 (75,0)	3 (42,9)
Specific features	—	He took it away. RV	"Snowman"	Globular Shadow	Variable

Note: l/s – pulmonary-vascular; RV — right ventricle.

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To assess the diagnostic significance of individual radiographic features in the verification of LGV, a ROC analysis was performed. The results are presented in Table 3.2.6.

Table 3.2.6. Diagnostic value of radiographic signs in the detection of high pulmonary hypertension

Sign	AUC	95% CI	Sensitivity (%)	Specificity (%)	Threshold
PNLA/tracheal diameter	0,86	0,76–0,93	79,2	84,8	≥1,50
CTI	0,74	0,63–0,83	72,1	68,4	≥0,63
Bulging of the second arch	0,71	0,60–0,80	87,5	55,3	Presence of a sign
Venous congestion	0,68	0,57–0,78	70,8	78,9	Presence of a sign
Interstitial edema	0,64	0,53–0,74	50,0	92,1	Presence of a sign

Note: AUC is the area under the ROC curve; PNLA — right descending pulmonary artery; CI is the confidence interval.

The ratio of the diameter of the PNLA to the diameter of the trachea (AUC=0.86) had the highest diagnostic value, which is consistent with the literature data on the high informative value of this indicator in LGV in children. The combination of three signs — cardiomegaly (CTI≥0.63) + protrusion of the second arch + expansion of the lung roots — provided a sensitivity of 91.7% and specificity of 73.7% in the diagnosis of high LGV.

Cross-expert agreement in the assessment of radiographic features was high: the Cohen kappa coefficient was κ=0.82 (95% CI: 0.74–0.89; $p<0.001$) for the assessment of cardiac shadow configuration, κ=0.79 (95% CI: 0.71–0.87; $p<0.001$) for the assessment of the pulmonary vascular pattern, and κ=0.88 (95% CI: 0.81–0.94; $p<0.001$) for the determination of cardiomegaly, indicating consistent reproducibility of the results.

Conclusion

Thus, chest X-ray examination in 123 patients with CHD complicated by high pulmonary hypertension revealed a characteristic set of signs, including cardiomegaly (80.5%), increased pulmonary vascular pattern (95.9%), dilation of

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the lung roots (91.1%), and bulging of the pulmonary artery arch (72.4%). A significant direct relationship was established between the degree of LGH and the values of the CTE ($r=0.47$; $p<0.001$) and the ratio of the diameter of the PNLA to the diameter of the trachea ($r=0.68$; $p<0.001$). The ratio of the diameter of the PNLA to the diameter of the trachea (AUC=0.86) had the highest diagnostic value, and the combination of three radiographic signs provided a sensitivity of 91.7% in detecting high LGH. Nosoa-specific patterns, such as "snowman" in TADLV, "egg on the side" in TMS, and spherical shadow in VKA, retain diagnostic significance in the context of concomitant LGV, although they can be modified by the severity of hemodynamic disorders. The phenomenon of decreased pulmonary pattern intensity in ultra-high LGV (AvDLA >70 mm Hg) is an unfavorable prognostic marker of irreversible pulmonary vasculopathy and requires immediate correction of treatment tactics.

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