Stages of arrythmogenic cardiomyopathy syndrome in children

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Arrhythmogenic cardiomyopathy determines clinical and prognostic arrhythmia value

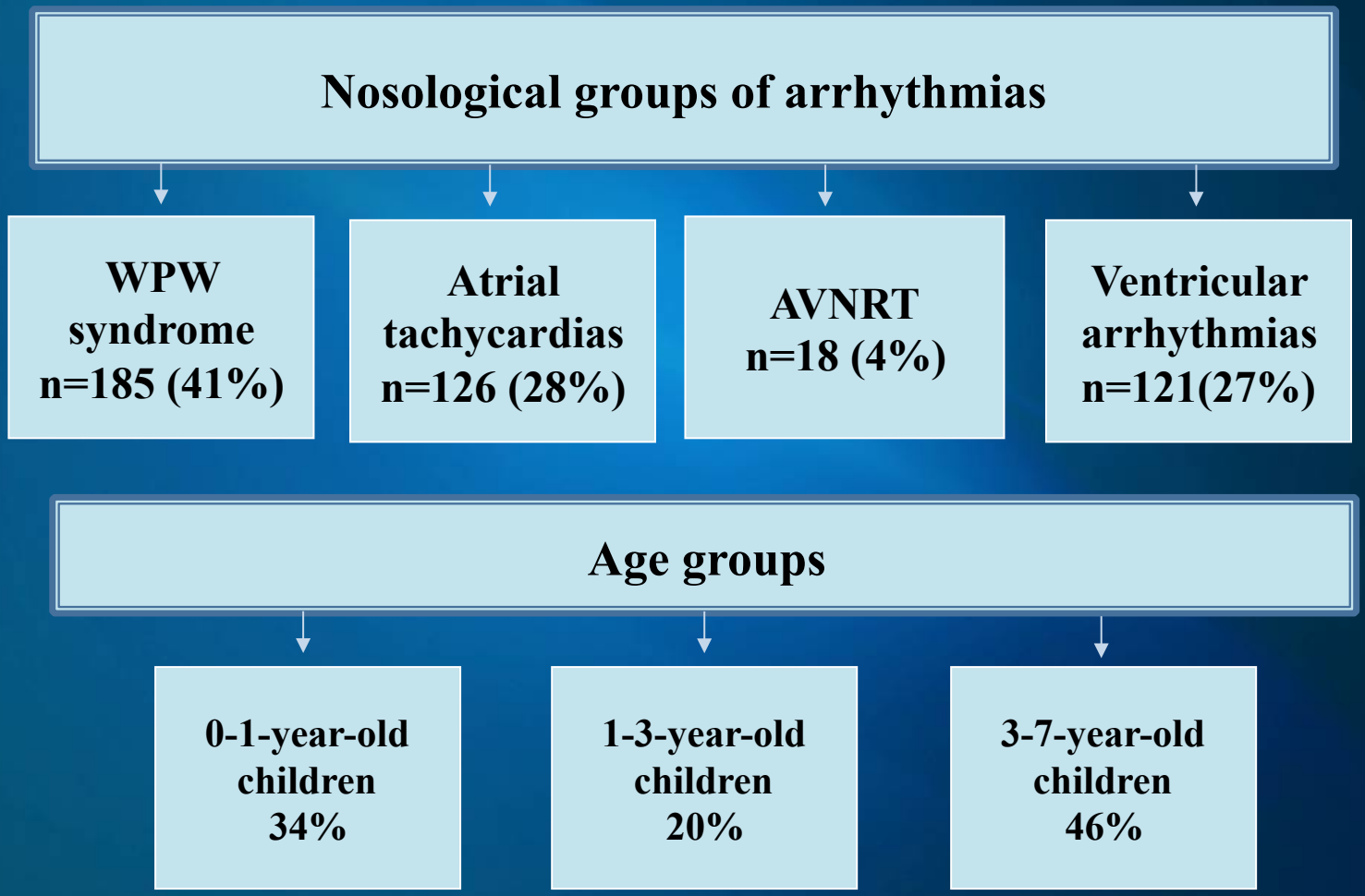
ACMP diagnostics criteria:

- Dilatation of heart chambers, decrease of LV contractile function
- Heart failure signs
- Reversibility of above-mentioned criteria after tachycardia resolution

Reasons for ACMP appearance

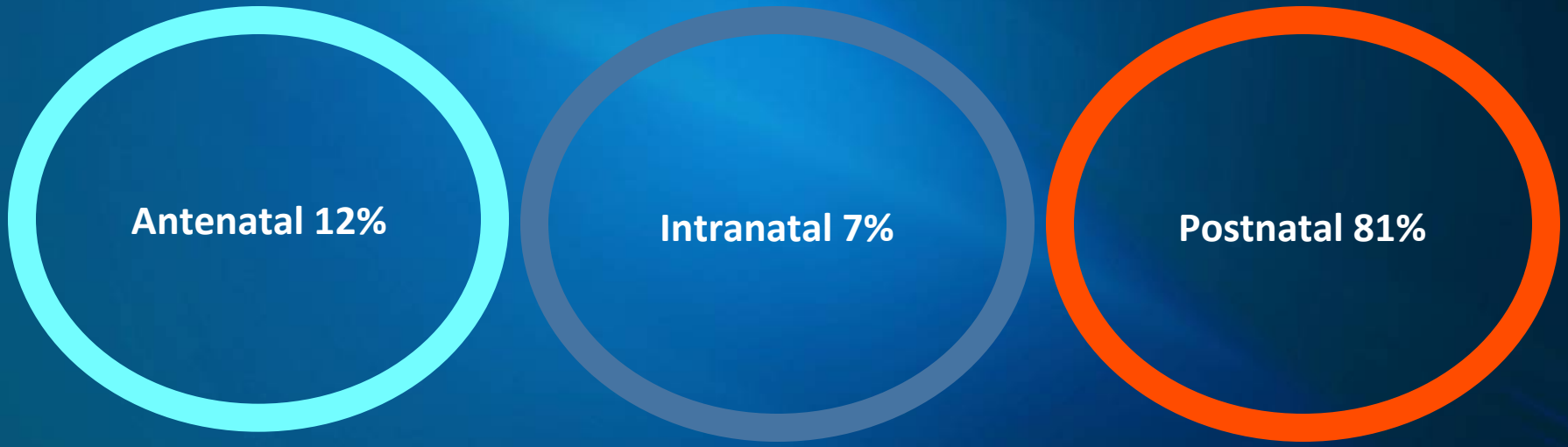
- Ectopic atrial tachycardia
- Ventricular tachycardia
- Ventricular arrhythmia with high ectopic activity
- WPW syndrome
- PJRT
- JET

Patients: 450 children with idiopathic arrhythmias aged 0 to 7 years (Me2,33(IQR: 0,33 - 5,0) were examined.



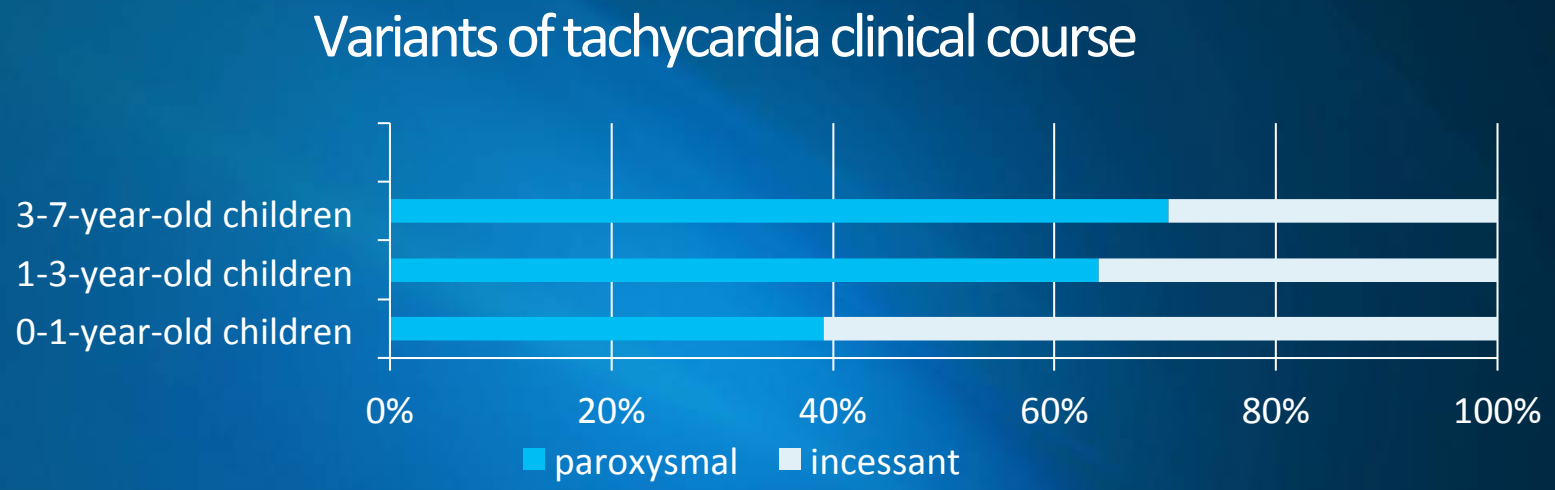
Arrhythmia manifestation period

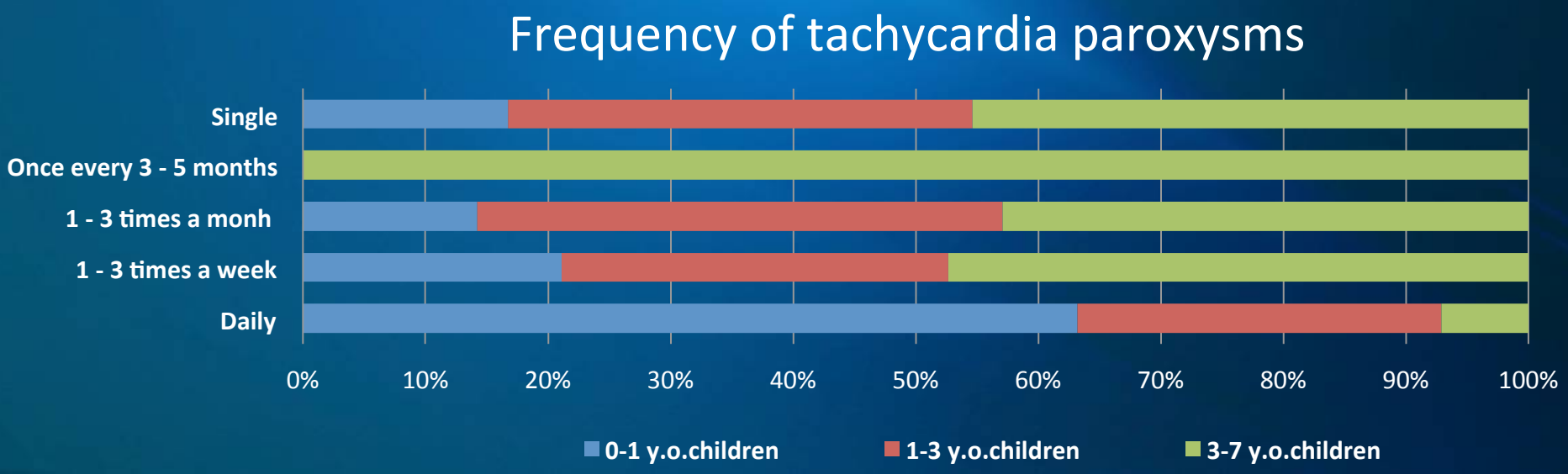
Period of the first arrhythmia manifestation was based on the recorded data of fetal Echo, auscultation, ECG, Holter.



The analysis of variants of tachycardia course in children of different age groups shown prevalence of paroxysmal course of tachycardia in children from 3 to 7 years old (p=0,001) and incessant one in children under one year old (p<0,001) .

Higher frequency of tachycardia paroxysms in children younger than one year in comparison with 1-3-year-old (p=0,028) and 3-7-year-old children (p=0,028)





Arrhythmogenic heart remodeling

Arrhythmogenic heart remodeling is specific either patients with chronic form of tachycardia or frequent tachycardia paroxysms.

Indices of deviation from age and anthropometric rates were assessed during individual Echo analysis.

- **Right atrial dilatation – 40.3%**
- **Left atrial dilatation – 35.4%**
- **Left ventricle dilatation – 35.4**
- **Right ventricle dilatation – 3.9%**
- **LV EF decrease – 49.2%**
- **LV spherification – 11.1%**
- **LV asynchronous contraction – 6.6%**
- **Enlargement of RV systolic pressure – 3.9%**
- **Mitral regurgitation – 18.8%**
- **Tricuspid regurgitation – 17.7%**

Significant prevalence of ACMP in older children in relation to younger children (p=0,002) was shown in assessment of Echo indices

Index	Age group			P intergroup	P paired		
	Under one year old	1-3 years old	3-7 years old		P 1-2	P 1-3	P2-3
LA volume,%	**98,3** (89,7-126,0)	**106,0** (88,3-132,0)	**126,5** (104,0-165,0)	**0,001**	0,400	**<0,001**	**0,020**
RA volume,%	**99,9** (80,7-137,0)	**112,0** (95,2-124,0)	**131,0** (116,0-151,0)	**<0,001**	0,373	**<0,001**	**<0,001**
EDV LV, %	**113** (84,1-131,4)	**117,5** (98,3-139,0	**112,5** (96,5-129,0)	0,488	-	-	-
EF LV	**66,0** (52,5-72,0)	**67,0** (60,0-70,0)	**65,0** (56,0-68,0)	0,708	-	-	-
RVSP	**25,0** (23,0-28,0)	**20,0** (19,0-22,5)	**23,0** (21,0-26,0)	**0,002**	**0,001**	**0,038**	**0,012**

Notes: LA volume,%, RA volume,% left atrium- and right atrium volume in % from age and anthropometric rates; EDV LV, % - end diastolic volume in % from age and anthropometric rates; EF LV - left ventricular ejection fraction; RVSP - right ventricular systolic blood pressure.

More over, EDV was less than 80% of individually prognosticated rates in 11 children. All those patients were under one year old

Heart failure (NYHA)

Significant difference in heart failure was shown between nosologies (p<0,001).

Heart failure develops more often in patients with atrial tachy (p=0,003).

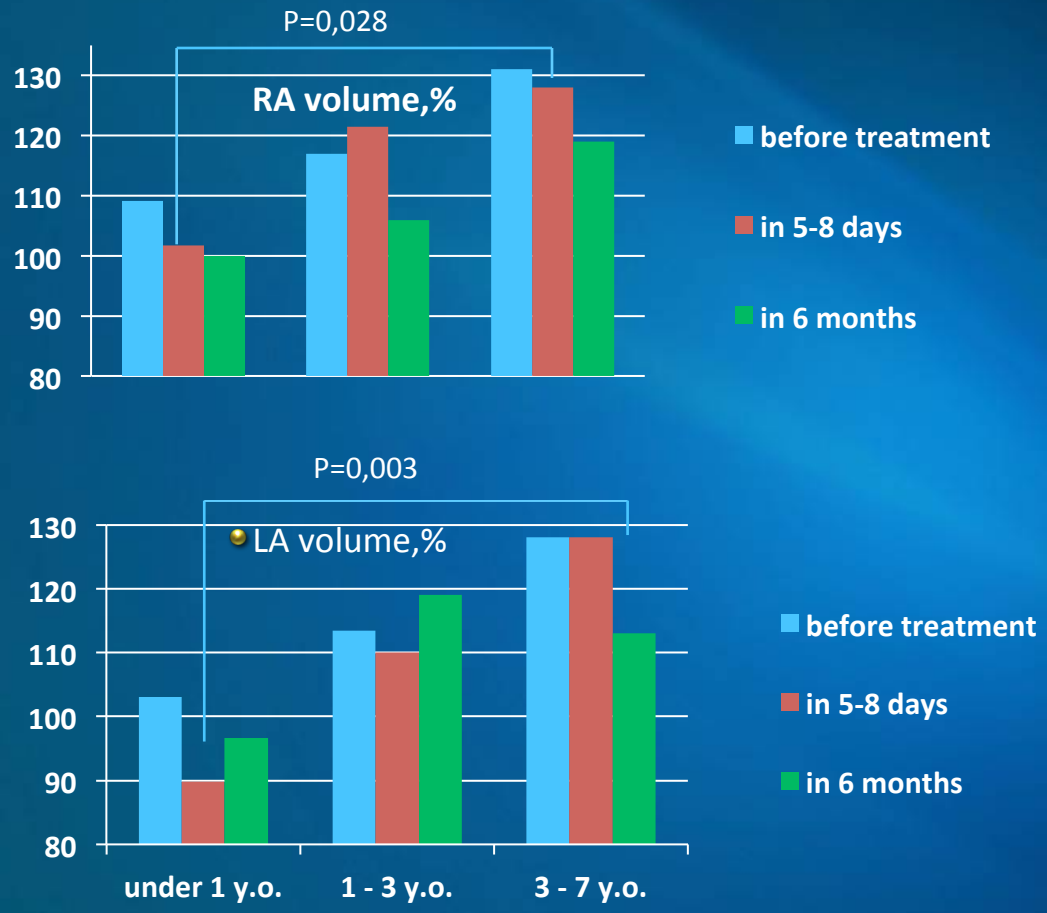
NYHA	Nosologic arrhythmias groups				Total
	WPW syndrome	Atrial tachycardias	AVNRT	Ventricular tachycardias	
I	61 (74,4%)	26 (45,5%)	7 (100%)	7 (53,8%)	101 (63,8%)
II	**13 (15,9%)**	**17 (39,9%)**	0	**5 (38,5%)**	35 (22,1%)
III	**8 (9,8%)**	**11 (20,0%)**	0	**1 (7,7%)**	20 (12,7%)
IV	0	**2 (3,6%)**	0	0	2 (1,3%)
Total	82 (100%)	56 (100%)	7 (100%)	13 (100%)	158 (100%)

Heart failure (NYHA)

NYHA	Age group			P intergroup	P paired		
	Under one year old	1-3 years old	3-7 years old		P 1-2	P 1-3	P2-3
I	38 (52,8%)	31 (83,8%)	70 (80,5%)	p=<0,001	p=0,004	p<0,001	p=0,934
II	16 (22,2%)	5 (13,5%)	14 (16,1%)				
III	17 (23,6%)	1 (2,7%)	2 (2,3%)				
IV	1 (1,4%)	0	1 (1,1%)				
Total	72 (100%)	37 (100%)	87 (100%)				

HF clinical manifestations significantly more often appeared in the group of children under one year old in comparison with children older age groups.

The important result of arrhythmias treatment by AAT and RFA is ACMP reverse remodeling



HF symptoms disappeared during first days after tachycardia resolution.

Reduction of echocardiographic signs of ACMP syndrome after arrhythmias resolution correlates with initial intracardiac hemodynamics.

Patients under one year old have been marked to have maximum benefit on intracardiac hemodynamics.

Conclusion

1. Arrhythmogenic heart remodeling occurred more frequently in children aged 3 to 7 years. Clinical signs of heart failure in infants with arrhythmias preceded structural changes and functional abnormalities on EchoCG. Normal sizes of cardiac chambers in the presence of high heart rates probably resulted in the diastolic dysfunction in infants. The factors leading to these hemodynamic changes were the high average daily heart rate, tendency of tachycardia to permanent and incessant course, and high frequency of paroxysmal tachycardia episodes in infants, short duration of history in infants.

2. To show arrhythmogenic remodeling and due to age and anthropometric patient inhomogeneity it is recommended to use deviance of the given indices from individually predicted anthropometric norms in percentage terms in order to assess dynamics of heart chambers volume.

3. Echocardiography in 6 and 12 months after arrhythmias resolution is recommended to assess ACMP reduction. Normalization of intracardiac hemodynamics depends on initial echocardiographic ACMP signs and duration of arrhythmia history.