A Retrospective Study In Children With Pulmonary Arterial Hypertension: A Single Center Experience

Kula S., Canbeyli F.H,,Atasayan V.,Tunağlu F.S., Oğuz A.D.

Gazi University Faculty of Medicine

Department of Pediatric Cardiology, Ankara, Turkey

INTRODUCTION

Pulmonary arterial hypertension (PAH)

- Heterogeneous
- Progressive
- Hard to manage
- Limited data in children

INTRODUCTION

- To determine the epidemiology, quality of life and management and effectiveness of the treatment of pediatric pulmonary arterial hypertension

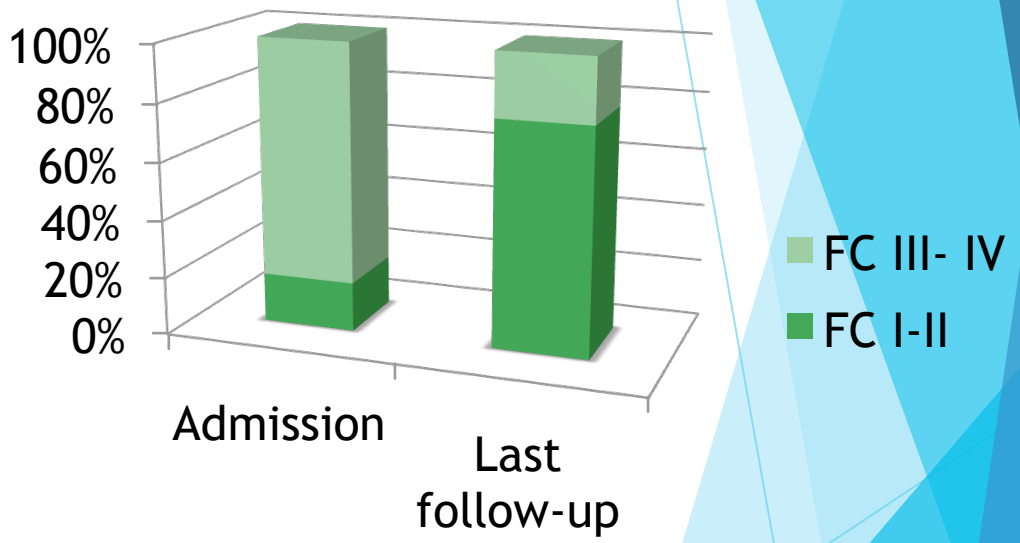
MATERIAL & METHOD

- Retrospective study
- Forty-one patients diagnosed with PAH and followed from February 2006 until October 2015
- Demographic characteristics, etiology, echocardiographic findings, 6-minute walking test, NYHA functional classification, catheterization findings, treatment methods, vasoreactivity test (VR), follow-up

RESULTS

Demographic Characteristics	n (%)
Female / Male	**21 (51.2) / 20 (48.8)**
Age	**83±68 months**
Etiology	
Congenital Heart Defects	**35 (85.4)**
VSD	12 (29.3)
ASD	1 (2.4)
ASD + VSD	7 (17)
ASD + absence of right PA	1 (2.4)
DORV + VSD	1 (2.4)
TGA + VSD	1 (2.4)
C-TGA + VSD	1 (2.4)
AVSD	2 (4.9)
TGA	2 (4.9)
TPVRA	1 (2.4)
Truncus arteriozus Type 1	1 (2.4)
Tricuspid atresia + ASD +VSD+PS	1 (2.4)
AP window	2 (4.9)
Diffuse pulmonary arterial hypoplasia	2 (4.9)
Primary PAH	**4 (9.8)**
Chronic Obstructive Lung Disease	**1 (2.4)**
Chronic Lung Disease	**1 (2.4)**
NYHA Functional Class	
I	-
II	**7 (17)**
III	**29 (71)**
IV	**5 (12)**

- Catheterization: 40/41 performed
- Statistically no difference between mPAP with catheterization and mPAP with echocardiography (65±20 mmHg vs 60±53.5 mmHg, p:0.268)
- VR test performed 24/40 (60%) and 39% positive
- Last follow up: Twenty-one of 29 patients with FC III in first admission, improved to FC II, 5 patients with FC IV improved to FC III, 7 patients with FC II improved to FC I
- Four patients died
- One and five year survival rates were 94% and 86%, respectively



	Admission	Last follow up	*p value*
6-MWT (m)	363 ±134	423 ±144	**0.001**
ProBNP (pg/ml)	282 ± 1514	176± 1686	**0.03**
Uric Acid (mg/dl)	4.2± 1.37	5.1± 2.04	**0.02**
AST (U/L)	30.3 ± 10.9	30.7 ± 21.04	0.11
ALT (U/L)	16.2 ± 6.3	17.2 ± 16.6	0.29
BUN (mg/dl)	12.6 ± 4.5	12.8 ± 3.8	0.51
Creatinine (mg/dl)	0.49 ± 0.25	0.49 ± 0.20	0.44
Hemoglobin (g/dl)	13.5 ± 2.54	14.5 ±2.9	**0.001**

	First Admission N (%)	Last follow up N (%)
No treatment	-	5 (%12.2)
Mono	23 (% 56)	11 (%26.8)
Dual	18 (% 44)	19 (%46.3)
Triple	-	6 (%14.6)
Bosentan	20 (%48.8)	10 (%24.4)
Sildenafil	2 (%4.9)	1 (%2.4)
İloprost	1 (%2.4)	-
Bosentan_+ sildenafil	7 (%17.1)	11 (%26.8)
Bosentan + iloprost	9 (%22)	6 (%14.6)
Sildenafil + iloprost	2 (%4.9)	2 (%4.9)
Bosentan + iloprost+ sildenafil	-	5 (%12.2)
Bosentan +iloprost+ tadalafil	-	1 (%2.4)

During the follow up, the combined therapy was increasingly prescribed (43.9% versus 66.9%). The mean time of adding second drug was 27 months

CONCLUSION

- At the experienced centers, the positive results on the survival rate and life quality of patients with PAH, obtained with the current treatment options
- As seen in this study, PAH is a progressive disease and the combined therapy is inevitable. Owing to this reason, the combined therapy should be considered in the early stage of the disease
- Only specific treatment not adequate
- **Check and support :** Anemia and heart failure, vaccination
- Multidisciplinary follow-up is essential