



Clinical, Laboratory , Echocardiographic and Hemodynamic Characteristics in Relation to Survival in Pulmonary Arterial Hypertension Associated with Congenital Heart Disease

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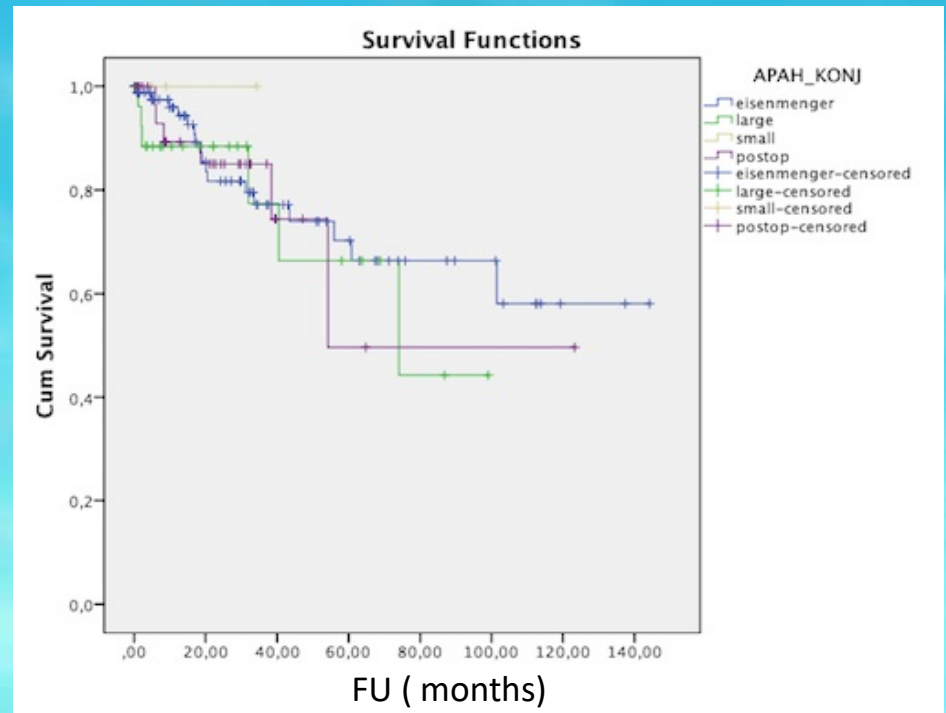
- Pulmonary arterial hypertension (**PAH**) associated with congenital heart disease (**APAHD-CHD**) has been classified as a subgroup of **PAH** that includes four different forms sharing a similar clinical and pathological findings.
- However, scientific interests have focused on the **Eisenmenger syndrome (ES)** while clinical characteristics, management strategy and predictors of the survival in **other subgroups** of patients (pts) with **APAHD-CHD** have remain to be determined.

- We aimed to investigate clinical, biochemical, neurohumoral, echocardiographic and hemodynamic predictors of the survival in 134 pts with **APAH-CHD** (Female 72 , age 40.7 ± 17.5 yrs) out of the 341 pts included into the **our single-center, prospective EUPHRATES (EvalUation of Pulmonary Hypertension Risk Factors AssociaTEd with Survival)** study.
- Subgroups of APAH-CHD were as follows ;
 - 1.ES (n=71),
 2. PAH associated with prevalent systemic-to-pulmonary shunts (n=37), and
 4. APAH which developed after defect correction (n=26). There was no pt with subgroup 3 .

- Baseline six-minute walk distance (**6MWD**) was 301.5 ± 127.8 m, and pulmonary arterial systolic and mean pressures (**PASP, PAMP**, mm Hg) were 99.6 ± 32.9 and 64.7 ± 22.85 and transpulmonary and diastolic pressure gradients (**TPG, DPG**, mm Hg) were 51.8 ± 22.9 and 29.6 ± 20.3 , respectively.
- Baseline pulmonary and systemic vascular resistance (**PVR, SVR**, Wood U) were 10.6 ± 9.3 and 21.8 ± 9.6 , respectively.
- **PVR/SVR ratio** was 0.52 ± 0.37 .
- **Targeted PAH treatments (TT)** were noted in 100 (75 %) pts.
- **Mean follow-up** was 30.9 ± 28 (0,5- 122,6) months.

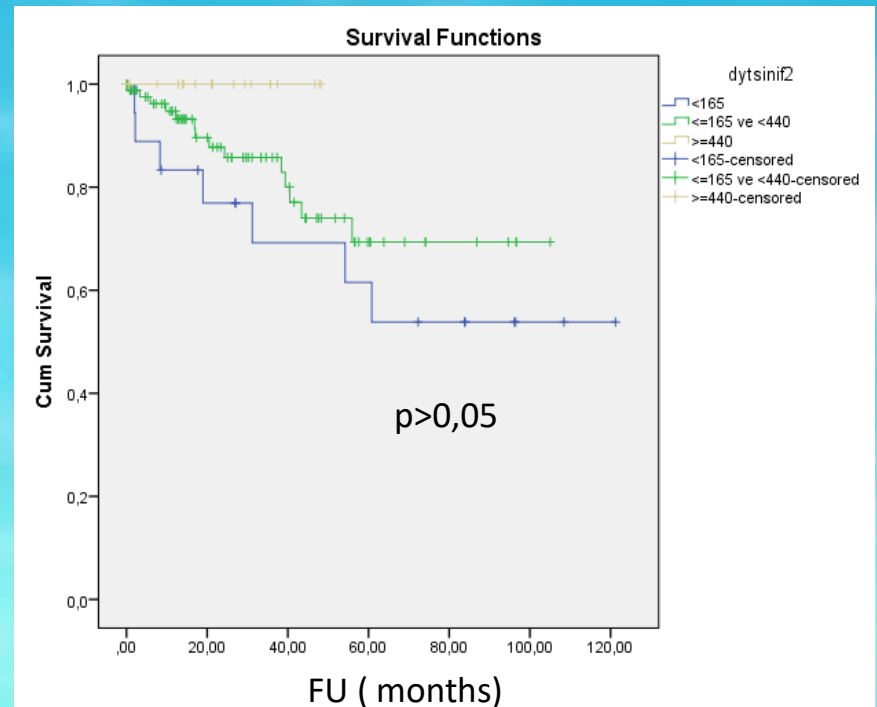
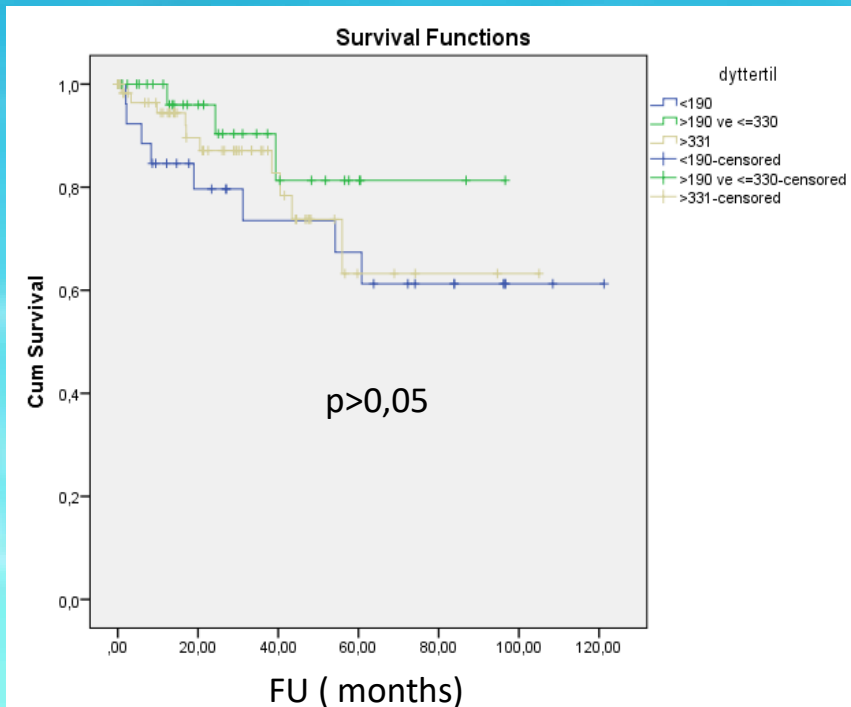
- The overall **APAH-CHD** group compared with idiopathic PAH (**IPAH**) and PAH-associated with connective tissue disease (**APAH-CTD**) groups had a better **8-year survival** (83 % vs 54,4 % and 61%, $p < 0,05$).

However, survival among three APAH-CHD subgroups were comparable ($p > 0,05$).

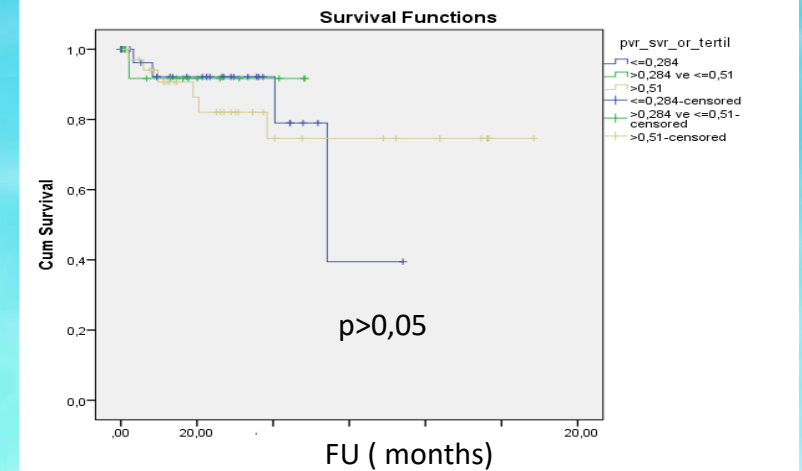
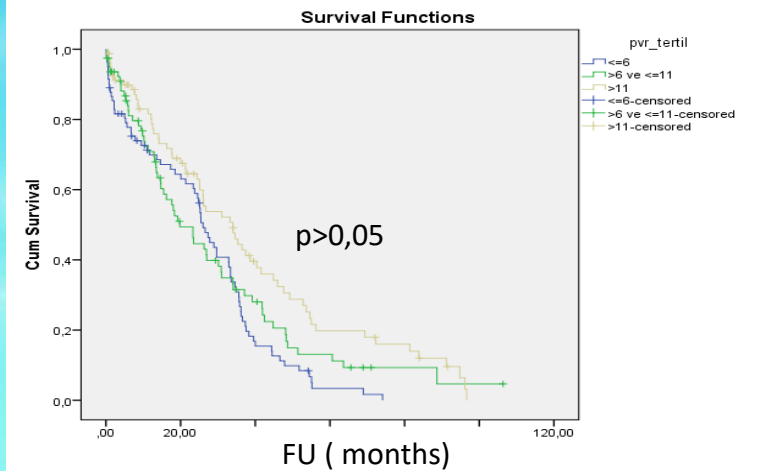
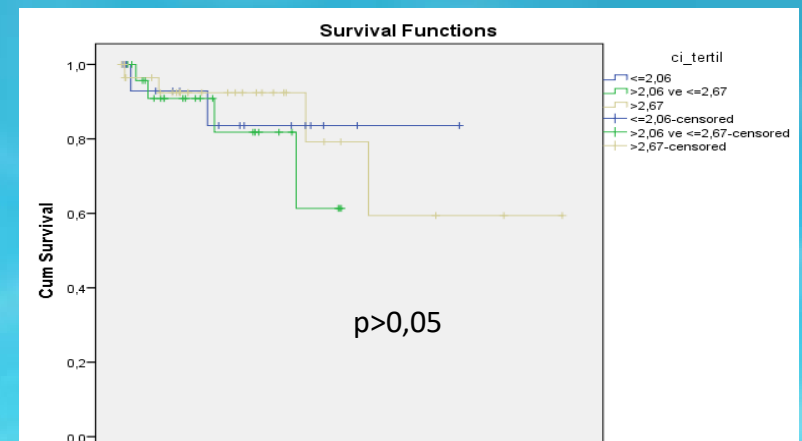
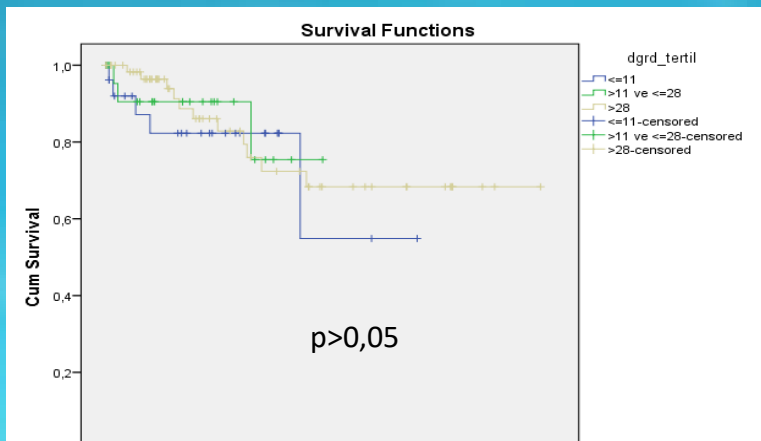


- The age, sex, functional class (FC), oxyhaemoglobin saturation %, C-reactive protein, (CRP), uric acid and brain natriuretic peptide (BNP) levels were **not related** with **survival difference** in pts with APAH-CHD($p > 0,05$).

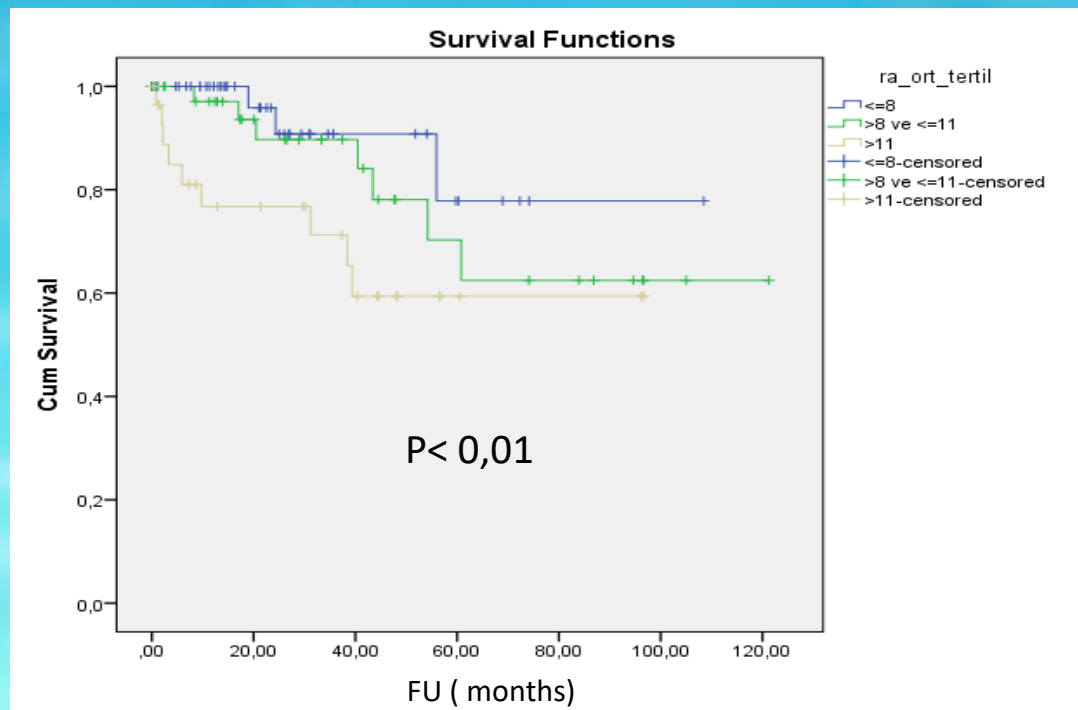
- **Neither tertile definition (<190, 190-330, > 331), nor pre-defined cut-off values of 6MWD (m) (<165, 165-440, > 440) related to survival difference in our APAH-CHD population ($p>0,05$).**



- Similarly, tertiles of TPG, DPG, CI, PVR and PVR/SVR ratio were not associated with survival difference ($p > 0,05$).



- However, presence of **pericardial effusion (PE)** ($p=0,0001$) and the highest tertile of **right atrial pressure (RAP, mm Hg)** (> 11 vs 9-11 and < 9 , $p<0.01$) were **associated a higher mortality in APAH-CHD.**



A second analysis in 170 pts with APAH-CHD out of the 481 PH pts.
Detailed results will be presented in ACC 17.

- **Pericardial effusion** ($p=0,008$),
- **IAS bulging to left side** ($p=0.003$),
- **Elevated RAP** ($p=0,002$),
- **WHO FC** (<0.001),
- **6MWD** ($p=0.026$),
- **CRP** ($p=0.002$),
- **BNP** ($p=0.025$),

but not other baseline measures, targeted treatment characteristics or clinical worsening episodes, were associated with **survival** in pts with APAH-CHD.

- The **APAH-CHD** pts seem to have a **better survival** as compared to those in **other PAH** subgroups.
- **Survival** was **comparable among** pts with **ES**, **APAH with prevalent left-to-right shunt**, and **APAH which developed after CHD defect correction**.
- The **PE, IAS bulging to left, FC, 6MWD, RAP, CRP** and **BNP** levels , **but not other** measures, **TT** or **clinical worsening** characteristics , **were associated with a higher mortality** in pts with **APAH-CHD**.

A photograph of two white doves sitting on a nest of twigs. They are facing each other and kissing. The background is a soft-focus view of a river with white water rapids.

***With Our Hopes for Peace Along the
EUPHRATES River,***

Thanks to;

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