



EBSTEIN'S ANOMALY AND TRICUSPID VALVE DYSPLASIA: PROGNOSIS AFTER DIAGNOSIS IN UTERO ?

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INTRODUCTION

- Ebstein's anomaly (EA) and Tricuspid valve dysplasia (TVD) are rare and complex congenital heart diseases
- When diagnosed before birth, they have a poor prognosis :
 - 45 % of intrauterine deaths
 - 35-40% neonatal deaths
 - frequent terminations
- Advances in prenatal diagnosis : early detection of these malformations
Very different anatomic and hemodynamic malformations : Prognosis is difficult
- Few and conflicting reports proposed prognostic criteria
- Aim of the study : evaluation of the different criteria
define reliable predictive factors for outcomes after fetal diagnosis

METHODS

- The study was retrospective, multicenter, performed in 7 cardiopediatric centers: Rouen, Tours, Amiens, Massy, Lille and Saint Denis de la Réunion.
- Between 1984 and June 2010, 37 fetuses were included all were followed-up. Each fetal echocardiography was reviewed
- We analyzed numbers of neonatal / fetal prognosis factors described in the literature (table1)
- Number of echocardiography differ for each fetus with an average of 1.7 [1 to 4]. The data presented were those with an acute prognosis independently of the chronology.
- 5 groups were classified according to their evolution: terminations, intrauterine deaths, neonatal deaths, late deaths (older than 2 years) and survivors.

RESULTS



	EA	TVD	TOTAL
Fetal death (hydrops)	4	1	5
Neonatal death	2	3	5
Late death	2	0	2
Still alive	10	4	14
Termination	8	3	11
Total	26	11	37

The mortality rate was 62 % with 47 % due to terminations.

5 anatomic pulmonary atresia: retrograde flow without PI throughout the pregnancy.

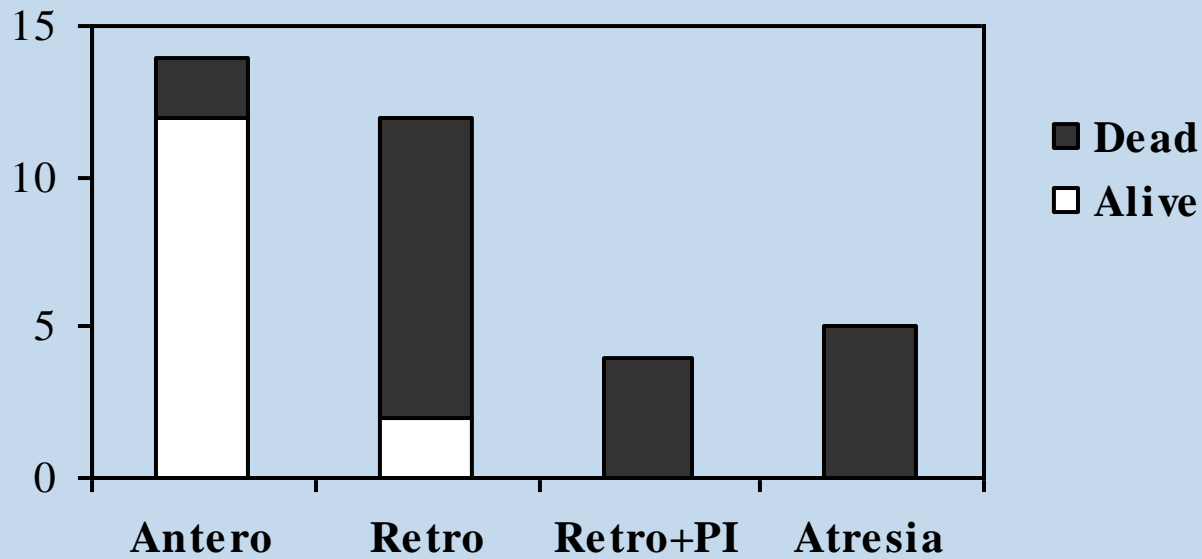
	Neonatal death	Late death	Survivors
Intensive care at birth	5	2	1
Surgical management	0	2 (Blalock)	1 (DAclosure)

		Survivors n = 14	Late death n = 2	Neonatal Death n = 5	Fetal death n = 5	p Survivor/ death
Gestation at diagnosis	< 27 weeks	8	1	4	5	ns
Cardio-thoracic ratio	< 0.65	4	1	0	0	ns
	0.65-0.75	6	1	3	1	
	>0.75	2	0	2	4	
RV/LV	< 1.5	9	1	5	2	ns
	1.5-2	5	1	0	2	
	> 2	0	0	0	1	
Celermajor index	<1	8	0	2	2	ns
	1-1.5	4	1	2	1	
	>1.5	2	1	1	2	
TI (grade)	Minim	3	0	0	0	0.04
	Moderate	6	2	0	0	
	Severe	5	0	5	5	
TI velocity	> 2.5m/s	5	0	2	1	-
	< 2.5 m/s	0	0	0	1	
Arantius flow	Normal	1	0	1	0	-
	Abnormal	0	0	1	2	
Arrhythmia	Prenatal	0	0	1	0	-
	Neonatal	2	0	0	0	
PA/Ao	< 0.5	0	1	0	2	0.04
	>0.5	9	3	1	2	

RESULTS

The major prognosis factor : THE PULMONARY ARTERY VALVE FLOW

$p < 0.05$

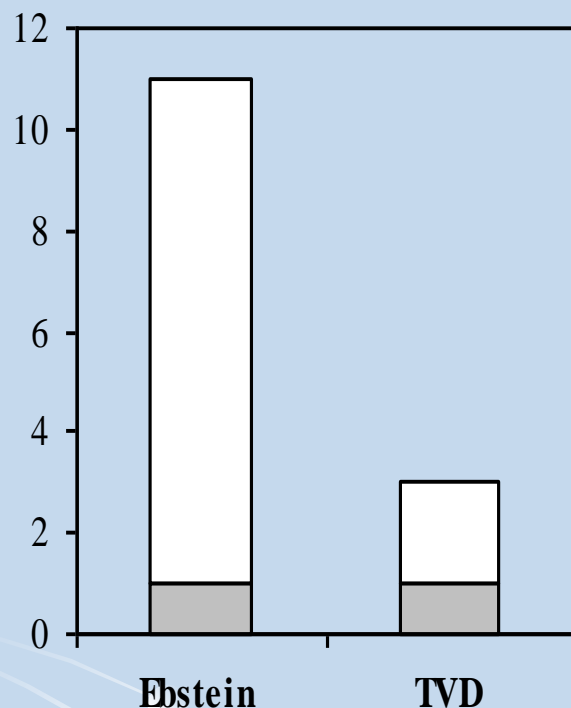


Retrograde PA flow had a PPV for death of 83 %

Associated with significant PI : functional pulmonary atresia / Pulmonary atresia PPV reach 100%

Anterograde PA flow predicted rather good outcomes with a PPV for survival of 86%

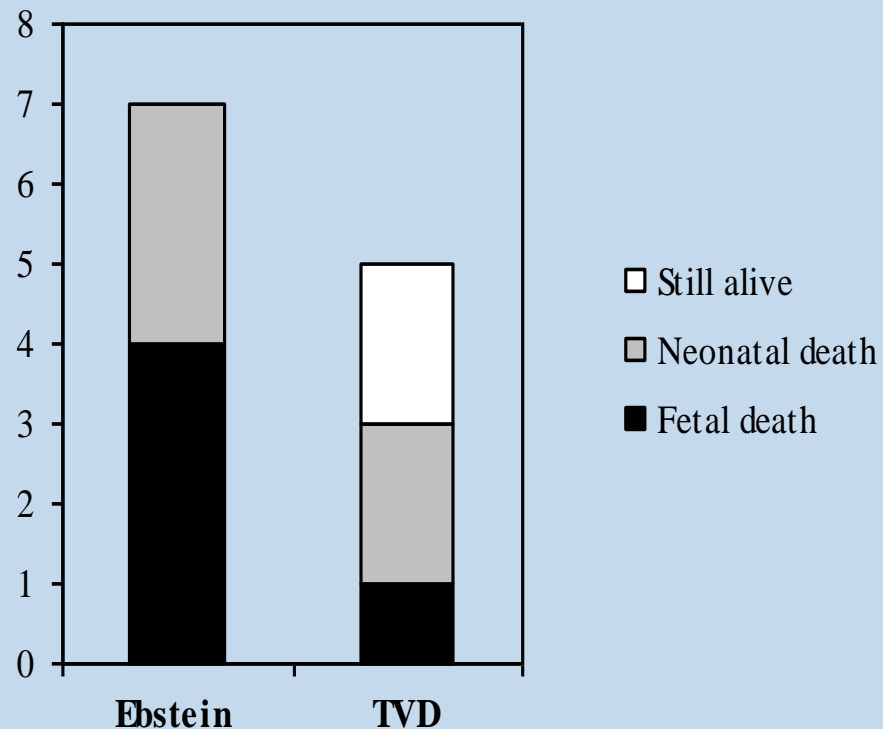
RESULTS



ANTERO GRADE PA Flow

EA : 1 died with an anterograde flow through a VSD

TVD : 1 died with a bidirectionnal flow due to high placental resistances



RETRO GRADE PA Flow

EA : 5 abnormal anterior leaflet / 3 massive TR or RV dysfunction

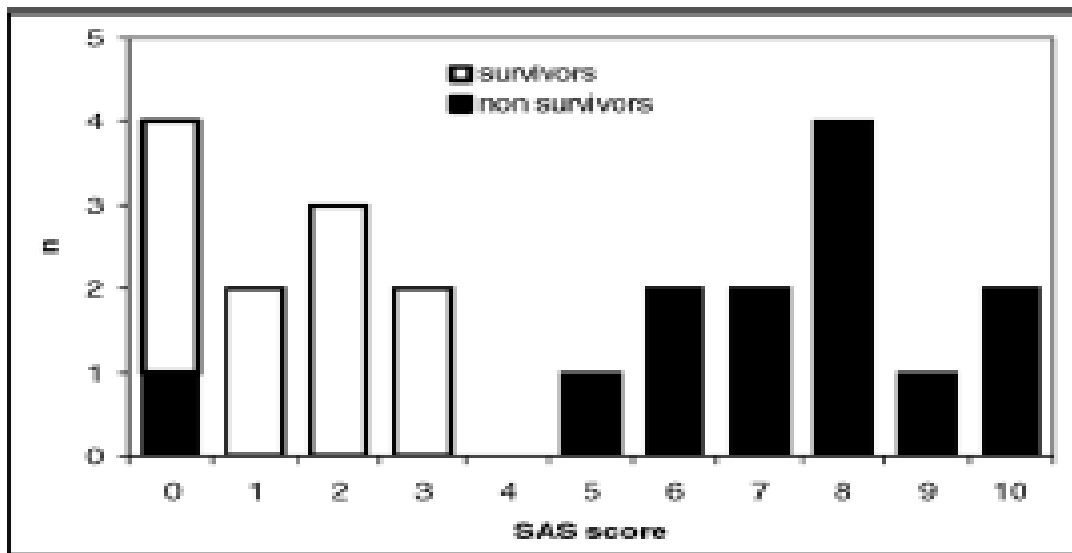
TVD: 3 recovered an anterograde flow at birth
2 died : prematurity/ large DA

RESULTS

The SAS score was a good prognosis factor $p < 0.05$



Variable	Weighting		
	0	1	2
Cardiothoracic ratio	<0.65	0.65–0.75	>0.75
Celermajer index	<1.0	1.0–1.5	>1.5
Pulmonary valve flow	Normal	Reduced	Absent
Duct flow	Anterograde	Both	Retrograde
Right–left ventricular Ratio	<1.5	1.5–2.0	>2.0



Anterograde
PA flow

Retrograde
PA flow

Retrograde
PA+ PI

Anatomic
Pulmonary
atresia (PA)

SAS score
 ≥ 5

SAS score
< 5

CONCLUSION



- EA and TVD : several hemodynamic and anatomic profiles
- The pulmonary valve flow is a **SIMPLE AND MAJOR PROGNOSTIC FACTOR** :
 - **RETROGRADE PA flow** : Sp for fetal /neonatal death 86 %
a PPV of death 100 % if associated to a massive PI or Pulmonary Atresia
 - **ANTEROGRADE PA flow** : Se of 86 % for good outcome if there is no VSD
- The follow up is fundamental : hemodynamic factors such as
 - Arrhythmia
 - Placental resistance
 - RV dysfunction / massive TR may deeply change the prognosis
- Valve anatomies is very important regarding the possibility of surgical repair