Figure 1. Prenatal Diagnosis Rate Over Time at a Single Obstetric Ultrasound Unit, All Major CHD

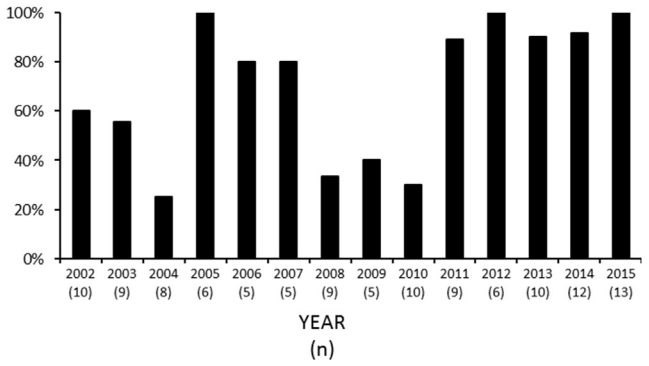
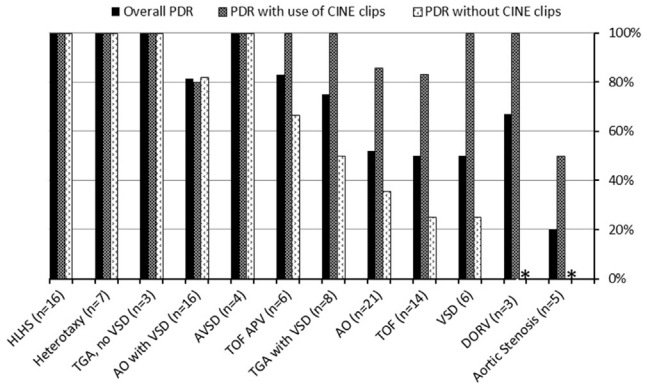


Figure 2. CINE Clip Use and Prenatal Diagnosis Rate (PDR) of Major CHD



207 Neonatal outcomes in Total Anomalous Pulmonary Venous Return (TAPVR): the role of prenatal diagnosis and pulmonary venous obstruction

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OBJECTIVE: To evaluate neonatal outcomes of Total Anomalous Pulmonary Venous Return (TAPVR) and identify fetal echocardiography (FE) findings associated with postnatal pulmonary venous obstruction (PPVO).

STUDY DESIGN: We retrospectively evaluated TAPVR cases seen at Children's Hospital Los Angeles from 2005-2014 and analyzed pre- and post-operative (op) outcomes based on prenatal diagnosis, PPVO, and heterotaxy syndrome. When available, FE was reviewed and measurements of pulmonary and vertical vein Dopplers were evaluated as prenatal predictors of PPVO. Univariate and multiple regression analyses and receiver operator characteristic (ROC) curve analyses were performed on SAS 9.4, p-valve <0.05 was considered significant.

RESULTS: Of 137 TAPVR cases, 16 (12%) were prenatally diagnosed, 82 (60%) had PPVO and 29 (21%) had heterotaxy. TAPVR repair was performed in 135 cases (Figure 1). Surgical mortality was 17.8% (24/135). Of the prenatally diagnosed patients, 62.5% also had heterotaxy syndrome. On univariate analysis, prenatal diagnosis and heterotaxy were both associated with higher mortality. Heterotaxy was also associated with longer length of intensive care stay, hospital stay, and pulmonary hypertension (sildenafil use). PPVO was associated with pre-op acidosis, shorter time to surgery and pulmonary hypertension (iNO use), but not with post-op mortality. The two patients who died prior to surgery had severe PPVO. On multiple regression analysis, heterotaxy was the only independent predictor of mortality in TAPVR (HR 4.2 (1.2-14), p=0.02). On univariate analysis, FE vertical vein Doppler indices predicted PPVO whereas pulmonary vein Doppler did not. ROC analysis indicated a vertical vein peak gradient > 1.6 mmHg predicted PPVO (100% sensitivity; 71% specificity).

CONCLUSION: TAPVR continues to have low prenatal diagnosis rates in the absence of heterotaxy syndrome, and severe neonatal morbidity and mortality regardless of PPVO. While PPVO patients had pre-op mortality and morbidity (lower pH and emergent surgery), heterotaxy syndrome was independently associated with significantly increased risk of mortality in TAPVR. Vertical vein gradient can help prenatally identify those at risk of PPVO and guide delivery management. Further efforts are needed to improve prenatal detection of isolated TAPVR.

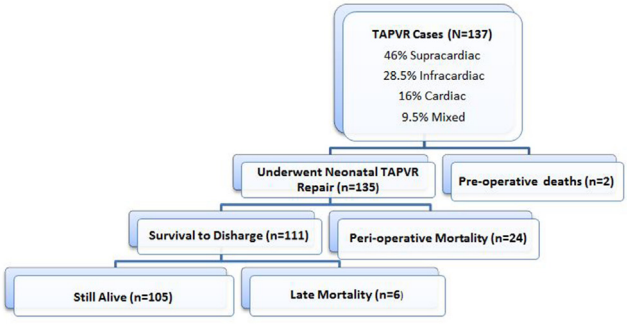

Figure 1: Of 137 patients enrolled, 135 underwent TAPVR repair. The 2 patients that died pre-operatively both had severe PPVO. Total mortality for the cohort was 32/137 (23.4%).

	TAPVR Outcomes	Prenatal (n=16) vs. Postnatal (n=121) Diagnosis	p-value	Obstructed (n=82) vs. Non-obstructed (n=55) Pulmonary Veins	p-value	Heterotaxy (n=29) vs. No Heterotaxy Syndrome (n=108)	p-value
Pre-op	GA at birth (weeks)	38.4±0.2 vs. 38.4±0.1	0.99	38.5±4.4 vs. 38.2±5.2	0.26	38.5±3.8 vs. 38.1±7.3	0.25
	Apgar score at 1 min	6.1±1.6 vs. 7.4±0.7	0.13	7.1±0.9 vs. 7.3±1.2	0.68	6.8±1.4 vs. 7.3±0.8	0.32
	Apgar score at 5 min	7.9±2.1 vs. 8.4±0.8	0.19	8.3±1.0 vs. 8.5±1.2	0.32	8.4±1.7 vs. 8.3±0.9	0.83
	Age at surgery (days)	5.1±1.4 vs. 6.3±0.6	0.31	5.1±0.6 vs. 7.8±1.1	<0.01	5.0±1.0 vs. 6.5±0.6	0.11
	iNO use	0% vs. 13.2%	0.02	15.9% vs. 5.5%	0.04	3.4% vs. 13.9%	0.14
	Lowest serum pH	7.2±1.9 vs. 7.2±0.7	0.95	7.16±0.8 vs. 7.24±1	<0.01	7.17±1.38 vs. 7.20±0.71	0.41
	Highest serum lactate	47.8±12.0 vs. 46.6±4.3	0.89	47.2±5.3 vs. 46.1±6.3	0.84	51.5±9.7 vs. 45.5±4.5	0.34
Post-op	ECMO use	7.4% vs. 19%	0.08	10% vs. 9%	0.84	10.3% vs. 9.3%	0.53
	ICU stay (days)	26.5±6.9 vs. 16.8±1.5	0.19	18.1±2.0 vs. 17.5±2.4	0.89	35.5±7.0 vs. 13.6±1.3	0.02
	Total hospital stay (days)	37.6±9.4 vs. 24.7±2.3	0.15	26.2±2.9 vs. 26.3±3.6	0.99	51.1±9.7 vs. 19.7±1.9	<0.01
	Acute renal failure	12.5% vs. 6.6%	0.07	7.3% vs.7.3%	0.89	13.8% vs. 5.6%	0.08
	iNO use	50% vs. 62.8%	0.14	75.6% vs. 40.0%	0.03	48.3% vs.64.8%	0.13
	Sildenafil use	12.5% vs. 12.4%	0.91	14.6% vs. 9.1%	0.34	24.1% vs. 9.3%	0.04
	Mortality	50.0% vs. 14.9%	<0.01	17.1% vs. 21.8%	0.63	46.9% vs. 10.5%	<0.01

Table 1: Pre-and post-operative outcomes were analyzed based on three categories: prenatal diagnosis, pulmonary venous obstruction, and heterotaxy syndrome.