

Initial Experience in Adults with Complex Congenital Heart Disease Treated with Sacubitril/Valsartan for Heart Failure

Tara Goodwin, Christopher Broda, MD; Angeline Opina, MD; Wilson Lam, MD; Peter Ermis, MD

Department of Pediatrics, Pediatric and Adult Congenital Cardiology, Baylor College of Medicine/Texas Children’s Hospital; Houston, TX



INTRODUCTION

- Over 90% of patients born with complex congenital heart disease are surviving to adulthood
- Adults with congenital heart disease (ACHD) are an emerging population and heart failure (HF) is a major cause of morbidity and mortality. Unfortunately, very few therapies have proven benefits for the ACHD patient with HF.
- Sacubitril/valsartan has proven effective treatment for HF with reduced ejection fraction in patients with acquired heart disease.
- There is little information available on the use of sacubitril/valsartan for treating HF in the ACHD population.

OBJECTIVE

- Evaluate the use of sacubitril/valsartan for the treatment of heart failure in adults with complex congenital heart disease.

METHODS

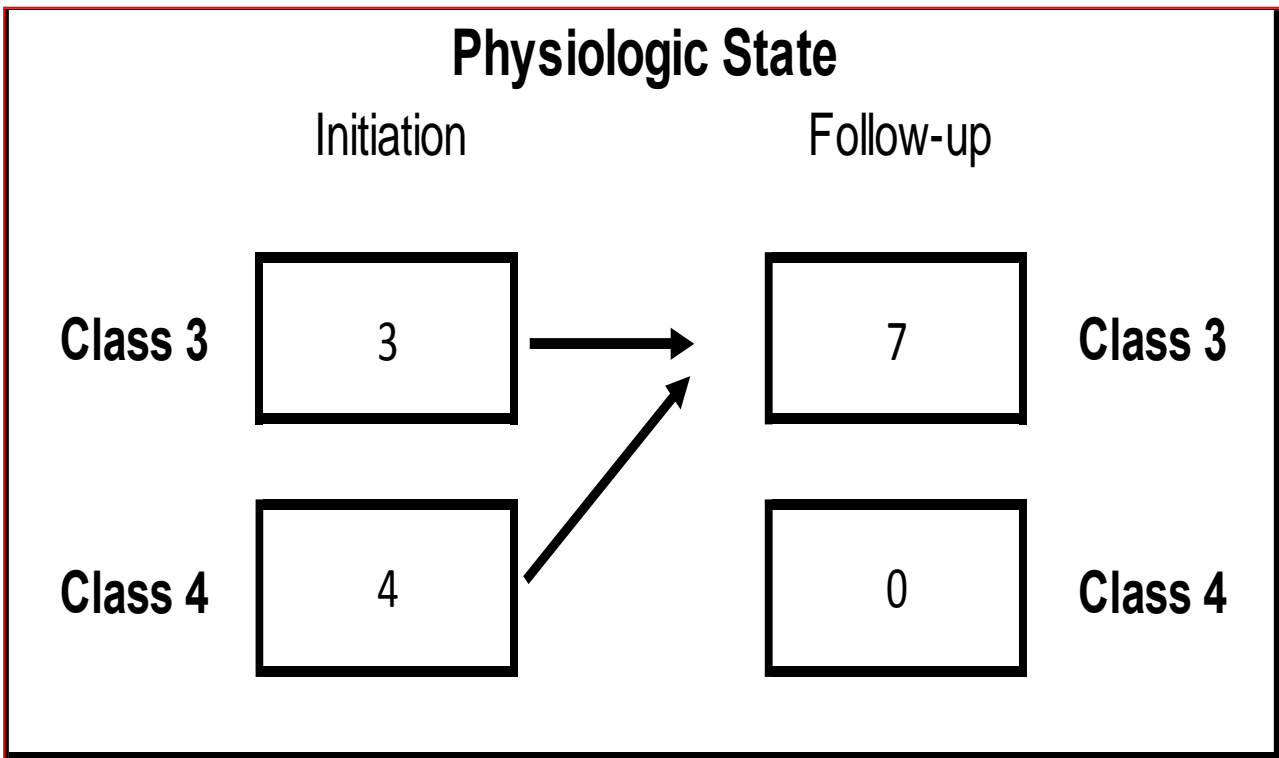
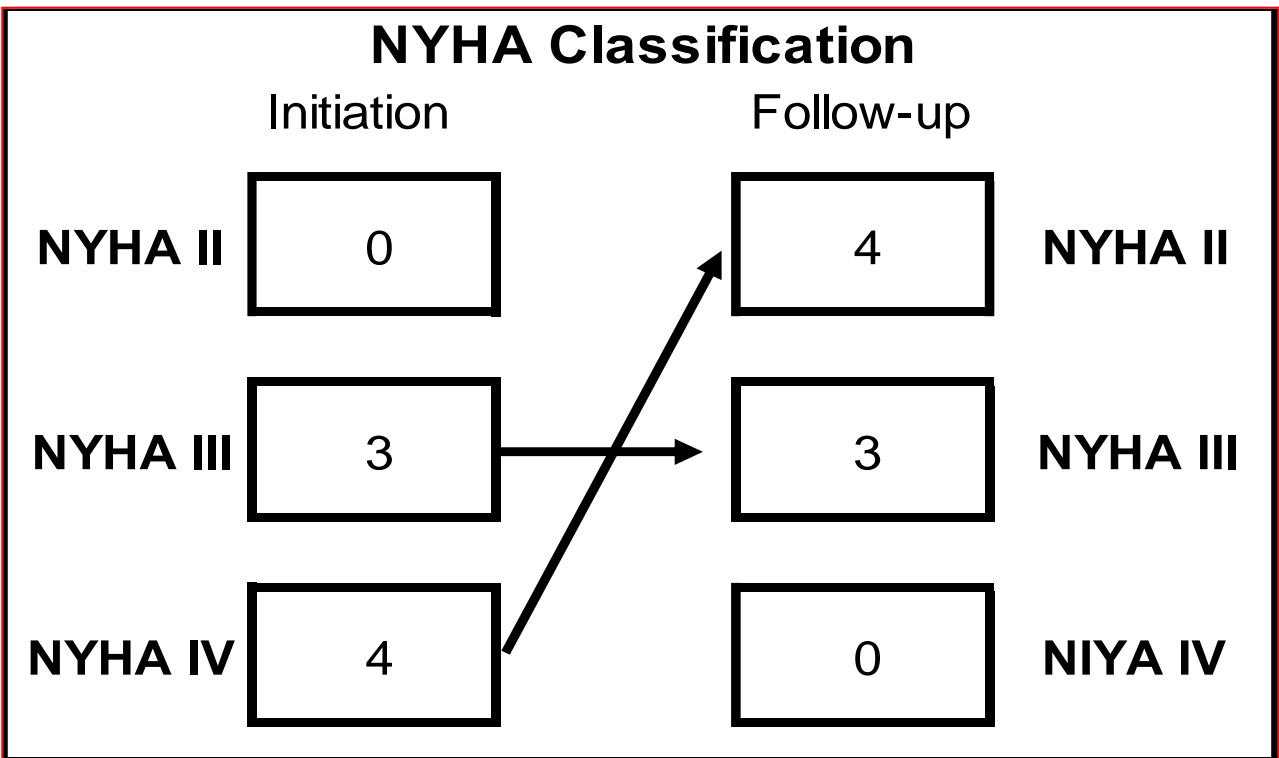
- Retrospective review at Texas Children’s Hospital of patients with great complexity congenital heart disease, >18 years of age who received sacubitril/valsartan for treatment of heart failure between 12/2017 and 10/2019
- Sacubitril/valsartan was initiated at time of heart failure exacerbation hospital admission or in outpatients with persistent HF symptoms despite medical therapy

RESULTS

Patient Demographics						
Patient	Age at Start of Sacubitril/Valsartan	Sex	Race	Diagnoses	Surgeries/Procedures	Pacemaker/ICD
1	55	M	White	D-TGA, VSD	atrial switch	Yes
2	47	F	White	D-TGA	atrial switch	yes
3	54	F	White	D-TGA	atrial switch	yes
4	33	M	White	D-TGA	atrial switch	no
5	38	F	Black	L-TGA, DORV, AV discordance	Fontan	no
6	24	M	Hispanic	DORV, MA	Fontan	yes
7	33	M	Hispanic	ccTGA, VSD, ASD		no
8	42	F	White	D-TGA	atrial switch	yes
M, male; F, female; D-TGA, dexto-transposition of the great arteries; VSD, ventricular septal defect; L-TGA, levo-transposition of the great arteries; DORV, double outlet right ventricle; AV, atrioventricular; MA, mitral atresia ; ccTGA, congenitally corrected transposition of the great arteries; ASD, atrial septal defect						

- Median age of 40.6 [IQR 33.3-52.8] years
- Median treatment duration 269 [IQR 102-473] days
- One patient has not returned for follow-up to evaluate functional class.
- One patient required dose reduction due to hypotension.
- 5/8 of patients required hospitalization for HF exacerbation after the initiation of sacubitril/valsartan.
- No patients experienced symptoms related to angioedema
- No patients have died or received advanced heart failure therapies (VAD or OHT) during the study period.

	Initiation Median [IQR], n(%)	Follow-up Median [IQR], n(%)
Dose of Sacubitril/valsartan		
• 24/26 mg	7 (87.5)	2 (25)
• 49/51 mg	1 (12.5)	4 (50)
• 97/103 mg	0 (0)	2 (25)
BNP	2517 [780-3181], n=5	1040, n=2
Creatinine (g/dL)	0.95 [0.71-1.68], n=6	0.96, n=3
GFR (ml/min/1.73m2)	81.5 [48.8-142.5], n=6	86, n=3
Potassium (g/dL)	4.6 [4.0-5.0], n=6	3.8, n=3
Weight (kg)	72.3 [58.5-82.425], n=6	73 [50.8-80.9], n=5
Heart rate (bpm)	88 [82-95]	76 [65-97], n=5
Mean arterial pressure (mmHg)	83 [75-95]	77 [72-82], n=5
Systemic ventricular function		
• Mild	1 (12.5)	0 (0)
• Moderate	2 (25)	0 (0)
• Severe	5 (62.5)	5 (100)
Subpulm ventricular function		
• Normal	2 (29)	0 (0)
• Mild	2(29)	2 (67)
• Moderate	1 (14)	0 (0)
• Severe	2 (29)	1 (33)



CONCLUSION

- Sacubitril/Valsartan was well tolerated in this patient group.
- Some patients experienced an improvement in NYHA Class.
- More studies on the use of sacubitril/valsartan in ACHD patients need to be done.
- A registry of outcomes across different centers in this patient population is needed.