

Medical Therapy for Systemic Right Ventricles: A Systematic Review (Part 1) for the 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease

Data Supplements

Table of Contents

Data Supplement 1. Literature Search Strategy: Beta Blockers, Angiotensin-Converting Enzyme Inhibitors, Aldosterone-Receptor Blockers, and Aldosterone Antagonists in Patients with Systemic Right Ventricles	2
Data Supplement 2. Full-Text Screened Articles.....	3
Data Supplement 3. Analysis of the Risk of Bias for PKVO ₂ and Systemic Ventricular Function Measured by CMR Imaging	5
Data Supplement 4. Methodological Quality Assessment Based on the Cochrane Collaboration Risk of Bias Tool.....	7
Data Supplement 5. Methodological Quality Assessment of Cohort Studies Based on the Newcastle-Ottawa Scale	8
References	9

Data Supplement 1. Literature Search Strategy: Beta Blockers, Angiotensin-Converting Enzyme Inhibitors, Aldosterone-Receptor Blockers, and Aldosterone Antagonist in Patients with Systemic Right Ventricles

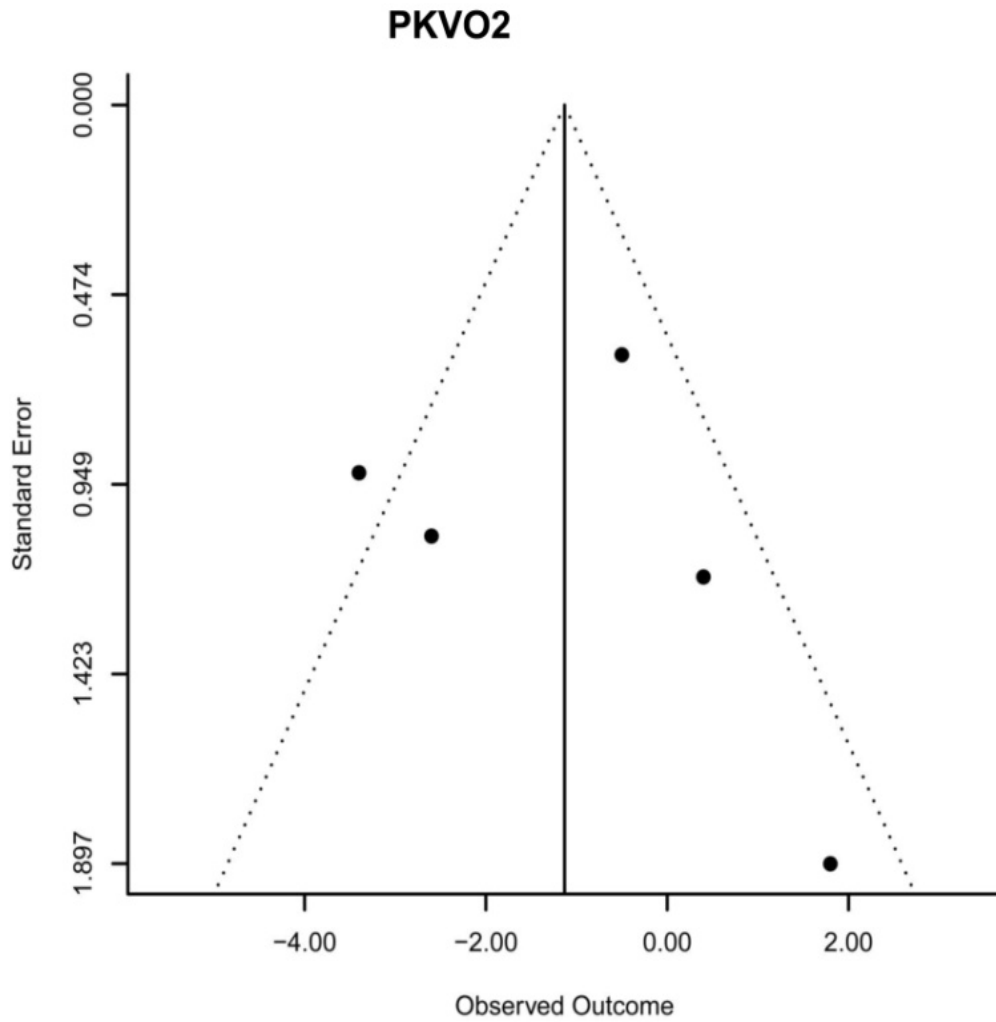
Search set	Query
#1	"heart single ventricle" OR "Transposition of Great Vessels"[Mesh] OR "Heart Ventricles/abnormalities"[MAJR] OR "univentricular heart" OR "double inlet left ventricle" OR "Tricuspid Atresia"[Mesh] OR "Norwood Procedures"[Mesh] OR "Fontan Procedure"[Mesh] OR "Pulmonary Atresia"[Mesh] OR "Transposition of the Great Vessels" OR "Dextro-Looped Transposition of the Great Arteries" OR "Congenitally corrected transposition of the great arteries" OR "Double outlet right ventricle" OR "Tricuspid Atresia" OR "Pulmonary Atresia" OR "Hypoplastic left heart syndrome" OR "Senning Operation" OR "Senning procedure" OR "Mustard operation" OR "Mustard procedure" OR "Atrial switch operation" OR "L-transposition of the great arteries" OR "D-transposition of the great arteries" OR "Pulmonary atresia" OR "Mitral Atresia" OR "Unbalanced atrioventricular canal defect" OR Heterotaxy OR "atrial baffle repair"
#2	"Angiotensin-Converting Enzyme Inhibitors"[Mesh] OR "Angiotensin-Converting Enzyme Inhibitors" [Pharmacological Action] OR "Adrenergic beta-Antagonists"[Mesh] OR "Adrenergic beta-Antagonists" [Pharmacological Action] OR "Beta blocker*" [title] OR "Beta blockade" [title] OR "ACE inhibitor*" [title] OR "beta adrenergic receptor blocking agent" OR "beta adrenergic receptor blocking" OR "metoprolol succinate" OR Metoprolol OR Bisoprolol OR carvedolol OR "Cardioselective beta 1 adrenergic blocker" OR "ace inhibitor" OR "angiotensin receptor blockers*" OR "angiotensin receptor antagonist" OR "angiotensin II type 1 receptor blocker" OR "angiotensin II receptor antagonist" OR valsartan OR losartan OR 'angiotensin converting enzyme inhibitors' OR 'lisinopril' OR enalapril OR benazepril OR captopril OR ramipril
#3	#1 AND #2
#4	#1 AND #2 AND Filters activated: Clinical Trial, Comparative Study, Controlled Clinical Trial, Meta-Analysis, Multicenter Study, Observational Study, Randomized Controlled Trial, Review, Systematic Reviews, Publication date from 1985/01/01 to 2014/12/31, Adolescent: 13-18 years, Adult: 19+ years, Young Adult: 19-24 years, Adult: 19-44 years, Middle Aged + Aged: 45+ years, Middle Aged: 45-64 years, Aged: 65+ years, 80 and over: 80+ years.

Data Supplement 2. Full-Text Screened Articles

1. Akutsu Y, Kaneko K, Kodama Y, et al. Sympathetic dysfunction of systemic right ventricle in congenitally corrected transposition of the great arteries. *Int J Cardiovasc Imaging*. 2010;26:483-4.
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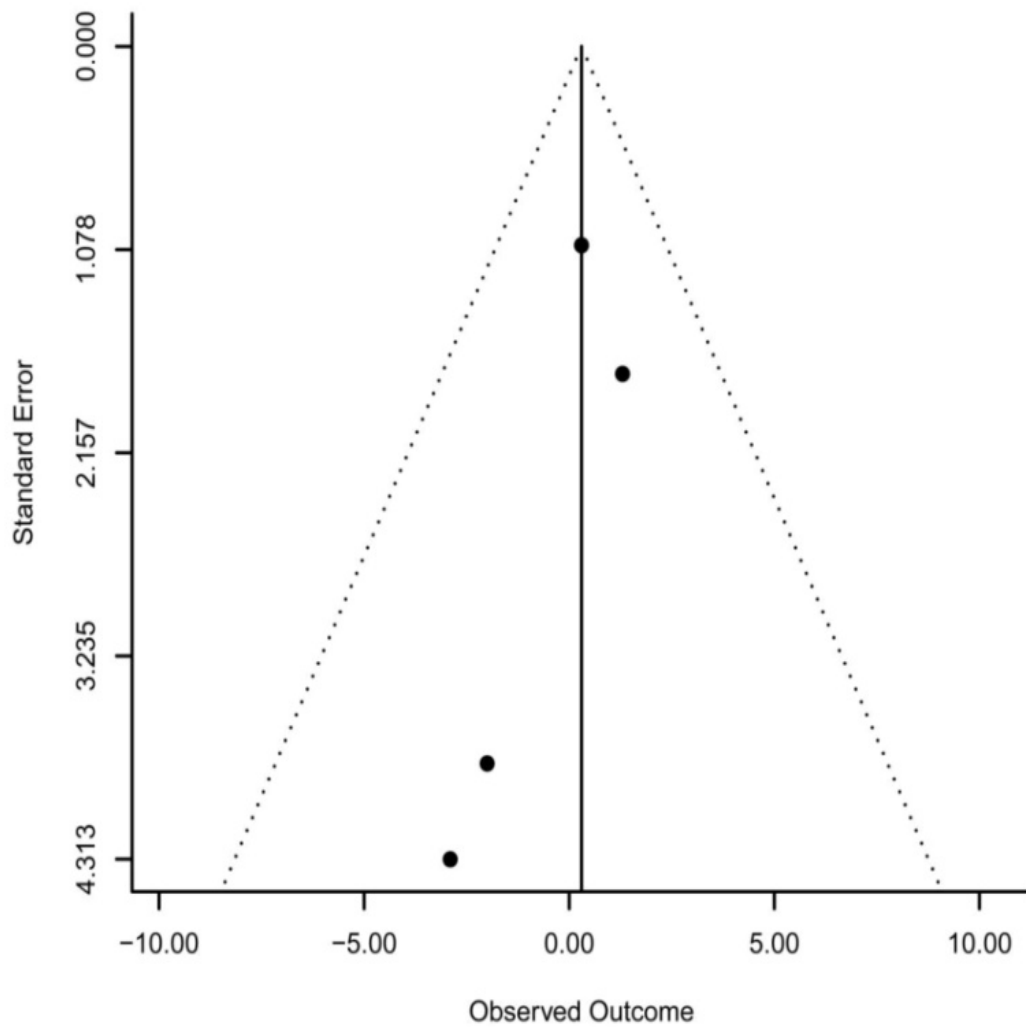
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Data Supplement 3. Analysis of the Risk of Bias for PKVO₂ and Systemic Ventricular Function Measured by CMR Imaging



Begg and Mazumdar's test (P-value): 0.333
Egger's test (P-value): 0.438

EFMRI



































Begg and Mazumdar's test (P-value): 0.483

Egger's test (P-value): 0.332

CMR indicates cardiovascular magnetic resonance; EF, ejection fraction; and PKVO₂ ventilatory equivalent of oxygen.

Data Supplement 4. Methodological Quality Assessment Based on the Cochrane Collaboration Risk of Bias Tool

	 Van Der Bomet al. (1) 23247302	 Dore et al. (2) 16216961	 Dos et al. (3) 23972966	 Therrien et al. (4) 18672299
Random sequence generation (selection bias)				
Allocation concealment (selection bias)				
Blinding of personnel and participants (performance bias)				
Blinding of outcome assessment (detection bias)				
Incomplete outcome data addressed (attrition bias)				
Free of selective reporting (reporting bias)				
Free of other bias				

Low risk of bias Unclear risk of bias High risk of bias

Data Supplement 5. Methodological Quality Assessment of Cohort Studies Based on the Newcastle-Ottawa Scale

	Hetcher et al. (5) 11230861	Tutarel et al. (6) 20843567	Giardini et al. (7) 21882492	Doughan et al. (8) 17317376	Boullal et al. (9) 20519056	Josephson et al. (10) 16835671
Representativeness*	0	0	0	0	0	0
Selection of non-exposed†	0	1	N/A	1	NA	N/A
Ascertainment of exposure‡	1	1	1	1	1	1
Outcome not present at study recruitment§	1	1	1	1	1	1
Comparability 	0	1	N/A	0	NA	N/A
Assessment of outcome¶	1	1	1	1	1	1
Follow-up long enough#	1	1	1	1	0	1
Rate of response**	1	1	N/A	1	N/A	N/A
Overall quality score††	5	7	4	6	3	4

*Representativeness of the exposed cohort (0,1); †Selection of the controls (0, 1, N/A); ‡Ascertainment of exposure (e.g., medical records) (0, 1, N/A); §Outcome of interest not present at start of study (0, 1); || Comparability of cases and controls for most important factor and additional factors (0, 1, 2, N/A); ¶Assessment of outcome (e.g., medical records) (0, 1, N/A); #Follow-up long enough, >4 months (0, 1, N/A); **Same nonresponse rate for both groups (0, 1, N/A); ††Total: minimum=1, maximum=9 stars. N/A indicates not applicable.

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4. Therrien J, Provost Y, Harrison J, et al. Effect of angiotensin receptor blockade on systemic right ventricular function and size: a small, randomized, placebo-controlled study. *Int J Cardiol*. 2008;129:187-92.
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