

**Known or Suspected Congenital Heart Disease in the Adult
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
1. Hoffman JI, Kaplan S. The incidence of congenital heart disease. <i>J Am Coll Cardiol.</i> 2002;39(12):1890-1900.	Review/Other-Dx	62 studies	To determine the reasons for the variability of the incidence of CHD, estimate its true value and provide data about the incidence of specific major forms of CHD.	The total incidence of CHD was related to the relative frequency of ventricular septal defects, the most common type of CHD. The incidences of individual major forms of CHD were determined from 44 studies. The incidence of CHD depends primarily on the number of small ventricular septal defects included in the series, and this number in turn depends upon how early the diagnosis is made. If major forms of CHD are stratified into trivial, moderate and severe categories, the variation in incidence depends mainly on the number of trivial lesions included. The incidence of moderate and severe forms of CHD is about 6/1,000 live births (19/1,000 live births if the potentially serious bicuspid aortic valve is included), and of all forms increases to 75/1,000 live births if tiny muscular ventricular septal defects present at birth and other trivial lesions are included.	4

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2. O'Leary JM, Siddiqi OK, de Ferranti S, Landzberg MJ, Opatowsky AR. The Changing Demographics of Congenital Heart Disease Hospitalizations in the United States, 1998 Through 2010. <i>JAMA</i> . 2013;309(10):984-986.	Review/Other-Dx	N/A	To analyze U.S. hospitalizations from 1998 through 2010 for children and ACHD.	The annual number of CHD hospitalizations among adults increased more quickly and consistently from 1998 through 2010 than pediatric hospitalizations. Adult admission volume was 87.8% higher during the second half of the study (n=622 084) compared with the first half (n=331 162), while pediatric admissions grew 32.8% (1 082 540 vs 815 471) ($P=0.001$ for interaction between age group and period). Admissions for simple defects changed 112.8%, unclassified defects changed 52.6%, and complex defects 52.8% between the eras among adults, compared with changes in simple defects (46.3%), unclassified defects (9.8%), and complex defects (32.4%) for children. Adults accounted for 36.5% (95% CI, 34.0%–38.9%) of CHD admissions in the latter era, up from 28.9% (95% CI, 26.6%–31.2%). The extent of medical comorbidity was greater among adults, though this increased significantly for both children and adults over the study period. Length of stay and inflation-adjusted hospital charges increased for both children and adults, but there was a modest decline in the proportion of admissions involving cardiac procedures among both groups.	4
3. Puranik R, Muthurangu V, Celermajer DS, Taylor AM. Congenital heart disease and multi-modality imaging. <i>Heart Lung Circ</i> . 2010;19(3):133-144.	Review/Other-Dx	N/A	To evaluate the role of imaging modalities in the management of subjects with CHD, particularly detailing recent developments in imaging techniques as they relate to the various CHD diagnoses commonly encountered in practice.	The recent improvements in noninvasive cross-sectional cardiovascular imaging modalities (MR and CT) have resulted in a change in our approach to the assessment and follow-up of patients with CHD. The precise role of these imaging modalities will evolve over time.	4
4. Marelli A, Gilboa S, Devine O, et al. Estimating the Congenital Heart Disease Population in the United States in 2010 – What Are the Numbers? <i>J Am Coll Cardiol</i> . 2012;59(13):E787.	Review/Other-Dx	N/A	To generate a contemporary estimate of the age and sex distribution of the CHD population in the U.S. through a combination of empirical and modeled data from the U.S. and Canada.	Estimated ranges for CHD prevalence and population size in the U.S. in year 2010 are presented. The estimated CHD population size in the U.S. in 2010 for males 0–17 is 444,000–468,000, females 0–17 is 482,000–508,000; males 18+ is 423,000–486,000, and females 18+ is 556,000–640,000 with a total estimation of 1,905,000–2,102,000.	4

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5. Moodie D. Adult congenital heart disease: past, present, and future. <i>Tex Heart Inst J.</i> 2011;38(6):705-706.	Review/Other-Dx	N/A	To briefly review the past, present, and future of ACHD.	The medical management of adult patients with CHD will continue to be a challenge, because the numbers of patients increase on a yearly basis while the expertise and personnel remain limited and focused upon a very few centers nationally.	4
6. Moons P, Bovijn L, Budts W, Belmans A, Gewillig M. Temporal trends in survival to adulthood among patients born with congenital heart disease from 1970 to 1992 in Belgium. <i>Circulation.</i> 2010;122(22):2264-2272.	Review/Other-Dx	7,497 patients	To investigate the proportion of CHD patients born between 1990 and 1992 who survived into adulthood and to compare their survival with that of CHD patients born in earlier eras and evaluate survival as a function of the type of heart defect.	Survival to 18 years of age in patients born between 1990 and 1992 was 88.6% (95% CI, 86.3% to 90.5%), which was significantly greater than that of patients born in previous decades ($P<0.0001$). For patients born between 1990 and 1992, survival into adulthood for those with mild heart defects was 98.0% (95% CI, 95.8% to 99.1%), whereas survival for those with moderate- and severe-complexity heart defects was 90.0% (95% CI, 86.8% to 92.5%) and 56.4% (95% CI, 47.4% to 64.5%), respectively. Analysis per heart defect confirmed these findings, demonstrating that patients with univentricular heart (49.1% [95% CI, 30.8% to 65.1%]) and hypoplastic left heart syndrome (7.5% [95% CI, 0.6% to 26.6%]) had the poorest survival rate.	4

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7. Beauchesne LM, Warnes CA, Connolly HM, Ammash NM, Tajik AJ, Danielson GK. Outcome of the unoperated adult who presents with congenitally corrected transposition of the great arteries. <i>J Am Coll Cardiol.</i> 2002;40(2):285-290.	Observational-Tx	44 patients	To determine the presentation and outcome of the unoperated adult with congenitally corrected transposition of the great arteries.	44 patients aged 20 to 79 years (mean, 44) were followed up to 144 months. In 29 (66%), the correct diagnosis was first made at age ≥ 18 years; the diagnosis was missed in 7 of these patients in a prior cardiology consultation, despite cardiac imaging. Systemic atrioventricular valve regurgitation (grade $\geq 3/4$) was noted in 26 patients (59%). 30 (68%) had surgical intervention, including systemic atrioventricular valve replacement in all, with no early mortality. Preoperatively, this subset had significant dysfunction of the systemic ventricle (EF, 40 +/- 10%), and most had advanced symptoms (25 with ability index $\geq 2/4$). In 16 (53%), systemic atrioventricular valve regurgitation $\geq 3/4$ and ventricular dysfunction had been documented for >6 months. The mean EF of the systemic ventricle decreased significantly postoperatively (34 +/- 11%, $P=0.006$). 4 patients (13%) eventually required cardiac transplantation. Poor preoperative EF of the systemic ventricle predicted eventual need for transplantation ($P=0.001$).	2
8. Romfh A, Pluchinotta FR, Porayette P, Valente AM, Sanders SP. Congenital Heart Defects in Adults : A Field Guide for Cardiologists. <i>J Clin Exp Cardiol.</i> 2012(Suppl 8).	Review/Other-Dx	N/A	To provide an overview of the anatomy of selected defects commonly seen in an adult congenital practice using pathology specimens and clinical imaging studies, and to describe the physiology, clinical presentation to the adult cardiologist, possible complications, treatment options, and outcomes.	In conclusion, the number of ACHD presenting for cardiology care is increasing each year. Successful surgical, interventional, and intensive care strategies have resulted in more living ACHD than children. Cardiologists caring for these patients must have a working knowledge of congenital heart defects, their treatment, complications and outcomes. This illustrated guide is presented as an aid for understanding and managing these complex and difficult patients.	4
9. Warnes CA, Liberthson R, Danielson GK, et al. Task force I: the changing profile of congenital heart disease in adult life. <i>J Am Coll Cardiol.</i> 2001;37(5):1170-1175.	Review/Other-Dx	N/A	To discuss the apparent prevalence in ACHD, disease patterns and diagnostic methods.	Role of echo and catheterization is discussed. Noninvasive methods and their increasing role mentioned.	4

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10. American College of Radiology. ACR Appropriateness Criteria®: Chest Pain Suggestive of Acute Coronary Syndrome. Available at: https://acsearch.acr.org/docs/69403/Narrative/ .	Review/Other-Dx	N/A	Evidence-based guidelines to assist referring physicians and other providers in making the most appropriate imaging or treatment decision for a specific clinical condition.	N/A	4
11. American College of Radiology. ACR Appropriateness Criteria®: Chronic Chest Pain — High Probability of Coronary Artery Disease . Available at: https://acsearch.acr.org/docs/69405/Narrative/ .	Review/Other-Dx	N/A	Evidence-based guidelines to assist referring physicians and other providers in making the most appropriate imaging or treatment decision for a specific clinical condition.	N/A	4
12. Laya BF, Goske MJ, Morrison S, et al. The accuracy of chest radiographs in the detection of congenital heart disease and in the diagnosis of specific congenital cardiac lesions. <i>Pediatr Radiol</i> . 2006;36(7):677-681.	Observational-Dx	281 patients	To determine the accuracy of radiologists in detecting CHD on the chest radiograph.	The average accuracy of the 5 readers in distinguishing normal from CHD patients was 78% (range of 72% to 82%). The overall measure of accuracy in distinguishing specific congenital cardiac lesions among 13 groups of patients was 71% (range of 63% to 79%).	3
13. Bhyravajhala S, Velam V, Polapragada NV, et al. Reliability of Doppler-Based Measurement of Pulmonary Vascular Resistance in Congenital Heart Disease with Left-to-Right Shunt Lesions. <i>Echocardiography</i> . 2015;32(6):1009-1014.	Observational-Dx	63 patients	To test the hypothesis that Doppler tricuspid regurgitation velocity/time-velocity integral of RV outflow tract ratio is a reliable noninvasive method for pulmonary vascular resistance assessment in patients with congenital shunt lesions across a wide range of pulmonary vascular resistance.	ROC curve plotted between the 2 methods showed good identity. Bland-Altman analysis showed excellent agreement between the 2 methods with negligible bias. ROC curves showed that pulmonary vascular resistance indexed echo was accurate in distinguishing different cutoff values of pulmonary vascular resistance in each of the 4 groups.	3

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14. Watson T, McCracken CE, Slesnick T, Kanaan U, Border WL, Sachdeva R. Quantitative Assessment of Ventricular Septal Contour for Estimation of Right Ventricular Pressure. <i>Echocardiography</i> . 2015.	Observational-Dx	108 subjects	To determine the relation between quantitative measures of septal flattening including the eccentricity index and a novel marker, the septal flattening angle with RV systolic pressure.	Of the 108 subjects, RV systolic pressure/systolic blood pressure was <50% in 77 and ≥50% in 31. In those with RV systolic pressure/systolic blood pressure ≥50%, the median septal flattening angle was significantly lower (7.4 degrees vs 22 degrees, $P<0.0001$), and the median eccentricity index was higher (1.61 vs 1.07, $P<0.0001$). Septal flattening angle and eccentricity index had a significant correlation with RV systolic pressure/systolic blood pressure ($r_s = -.70$ and 0.61 , respectively). AUC was higher for septal flattening angle compared to eccentricity index (0.92 and 0.85, respectively). The sensitivity and specificity of septal flattening angle for predicting an RV systolic pressure/systolic blood pressure ≥50% using a cut point of 16 degrees was 84% and 95% and for an eccentricity index cut point of 1.35 was 74.2% and 96.1%, respectively.	3
15. Babu-Narayan SV, Giannakoulas G, Valente AM, Li W, Gatzoulis MA. Imaging of congenital heart disease in adults. <i>Eur Heart J</i> . 2015.	Review/Other-Dx	N/A	To review the imaging of CHD in adults.	As a result ACHD, a heterogeneous population, benefit from appropriate application of multiple imaging modalities matched with tertiary ACHD expertise.	4

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16. Selly JB, Iriart X, Roubertie F, et al. Multivariable assessment of the right ventricle by echocardiography in patients with repaired tetralogy of Fallot undergoing pulmonary valve replacement: a comparative study with magnetic resonance imaging. <i>Arch Cardiovasc Dis.</i> 2015;108(1):5-15.	Observational-Dx	26 patients	To evaluate the accuracy of conventional echocardiographic variables and real-time 3D echocardiography in assessing RV volumes and function compared with MRI, in adult patients with repaired tetralogy of Fallot and referred for pulmonary valve replacement.	Correlations between conventional variables and MRI were absent or poor when assessing RV EF, except for fractional area of change ($r=0.70$, $P<0.01$ before pulmonary valve replacement; $r=0.68$, $P<0.01$ after pulmonary valve replacement) and real-time 3D echocardiography ($r=0.96$, $P<0.01$ before pulmonary valve replacement; $r=0.98$, $P<0.01$ after pulmonary valve replacement). The RV volume correlation between real-time 3D echocardiography and MRI was excellent before and after surgery for RV end-diastolic volume ($r=0.88$, $P<0.01$ and $r=0.91$, $P<0.01$, respectively) and RV end-systolic volume ($r=0.92$, $P<0.01$ and $r=0.95$, $P<0.01$, respectively). The accuracy of these indices, as a diagnostic test for impaired RV (<45%), was good: Youden's indexes varied from 0.47 to 0.89; AUCs before and after pulmonary valve replacement were 0.86 and 0.81 for fractional area of change and 0.98 and 0.97 for real-time 3D echocardiography, respectively.	2
17. Lam YY, Yu CM, Zhang Q, Yan BP, Yip GW. Enhanced detection of patent foramen ovale by systematic transthoracic saline contrast echocardiography. <i>Int J Cardiol.</i> 2011;152(1):24-27.	Observational-Dx	112 patients	To evaluate the effectiveness of transthoracic saline contrast echocardiography in detecting patent foramen ovale.	TEE identified patent foramen ovale in 45% of patients. The sensitivities of transthoracic saline contrast echocardiography in detecting patent foramen ovale at rest, during strain and release of Valsalva maneuver, and coughing were 12.0%, 38.0%, 80.0% and 94.0% respectively (each $P<0.05$ when compared to previous stage). Specificities were similar and >95% for all stages. Moreover, the release phase of the maneuver improved the diagnostic accuracy [defined as (number of true positives+true negatives) divided by total in sample] with incremental value over the preceding strain phase (89.2 vs 70.5%, $P<0.001$).	2

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18. Salehian O, Schwerzmann M, Merchant N, Webb GD, Siu SC, Therrien J. Assessment of systemic right ventricular function in patients with transposition of the great arteries using the myocardial performance index: comparison with cardiac magnetic resonance imaging. <i>Circulation</i> . 2004;110(20):3229-3233.	Observational-Dx	29 total patients: 11 with congenitally corrected transposition of the great arteries, 18 with surgically corrected transposition of the great arteries	To compare myocardial performance index and CMR in the assessment of systemic RV function in patients with transposition of the great arteries.	MRI successfully assessed RV function and was found to be in good agreement with echocardiography, with echocardiography having much wider interobserver variability. Myocardial performance index can be used in patients with systemic RVs to assess global function and to estimate an EF with good accuracy.	3
19. Kutty S, Colen T, Thompson RB, et al. Tricuspid regurgitation in hypoplastic left heart syndrome: mechanistic insights from 3-dimensional echocardiography and relationship with outcomes. <i>Circ Cardiovasc Imaging</i> . 2014;7(5):765-772.	Review/Other-Dx	70 patients	To test the following hypotheses: 1) patients with hypoplastic left heart syndrome who develop significant tricuspid regurgitation or require tricuspid valve surgery in the medium term have detectable tricuspid valve abnormalities by 3D echocardiography prestage 1 palliation and 2) tricuspid regurgitation is associated with reduced survival and increased tricuspid valve intervention.	Of 70 patients, 62 (88.6%) had mild or less tricuspid regurgitation and 8 (11.4%) had moderate or greater tricuspid regurgitation prestage 1. Prestage 1 tethering volume correlated to leaflet area ($r=0.736$; $P<0.001$), annulus area ($r=0.651$; $P<0.001$), RV end-diastolic area ($r=0.347$; $P=0.003$), fractional area change ($r=-0.387$; $P<0.001$), and tricuspid regurgitation grade ($r=0.447$; $P<0.001$). At follow-up, 46 (65.7%) had mild or less tricuspid regurgitation (group A) and 24 (34.3%) had moderate or greater tricuspid regurgitation (group B). Prestage 1 3D echocardiography showed greater tricuspid valve tethering volume and flatter annulus in group B. Survival was better in group A.	4

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20. Khoo NS, Young A, Occlshaw C, Cowan B, Zeng IS, Gentles TL. Assessments of right ventricular volume and function using three-dimensional echocardiography in older children and adults with congenital heart disease: comparison with cardiac magnetic resonance imaging. <i>J Am Soc Echocardiogr.</i> 2009;22(11):1279-1288.	Observational-Dx	28 of 54 consecutive patients	To assess RV volume and function using 3D echocardiography in older children and ACHD and to compare with CMR.	There was no significant difference in EF measurements between 3D echocardiography methods and MRI except for automated border detection (-2.6 +/- 6, P=.03). The mean analysis time for automated border detection was 5 minutes, compared with 19 minutes for manual tracing (P<.0001). Approximately half the patients with CHD had adequate 3D echocardiographic images. 3D echocardiography accurately estimated EF but underestimated volume, particularly when the RV was dilated. Automated border detection minimally underestimated EF but offered a significant reduction in analysis time.	1
21. Cheng TO, Xie MX, Wang XF, Wang Y, Lu Q. Real-time 3-dimensional echocardiography in assessing atrial and ventricular septal defects: an echocardiographic-surgical correlative study. <i>Am Heart J.</i> 2004;148(6):1091-1095.	Observational-Dx	38 patients with ASD and/or ventricular septal defect	To determine feasibility of real-time 3D echocardiography for evaluation of ASD and ventricular septal defect.	Real-time 3D echocardiography was feasible and provided additional information over traditional 2D echocardiography. However, the real-time 3D echocardiography images from 14 subjects (27%) were excluded because of inadequate quality from morbid obesity, narrow intercostals spaces, and severe pulmonary emphysema.	2
22. Crean AM, Maredia N, Ballard G, et al. 3D Echo systematically underestimates right ventricular volumes compared to cardiovascular magnetic resonance in adult congenital heart disease patients with moderate or severe RV dilatation. <i>J Cardiovasc Magn Reson.</i> 2011;13:78.	Observational-Dx	25 patients	To compare volumetric measurements of the RV in 25 patients with adult CHD using both CMR and 3D echocardiography.	Bland-Altman analysis of the 25 patients demonstrated that for both RV end-diastolic volumes and RV end-systolic volumes, there was a significant and systematic underestimation of volume by 3D echo compared to CMR. This bias led to a mean underestimation of RV end-diastolic volumes by -34% (95% CI, -91% to + 23%). The degree of underestimation was more marked for RV end-systolic volumes with a bias of -42% (95% CI, -117% to + 32%). There was also a tendency to overestimate RV EF by 3D echo with a bias of approximately 13% (95% CI, -52% to +27%).	3

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23. Diller GP, Radojevic J, Kempny A, et al. Systemic right ventricular longitudinal strain is reduced in adults with transposition of the great arteries, relates to subpulmonary ventricular function, and predicts adverse clinical outcome. <i>Am Heart J.</i> 2012;163(5):859-866.	Observational-Dx	129 patients and 38 healthy subjects	To investigate the value of indices of myocardial deformation on speckle-tracking echocardiography for quantifying ventricular function and their potential role in assessing ventricular-ventricular interaction and outcome in patients with a systemic RV.	Systemic ventricular longitudinal 2D peak systolic strain was significantly reduced compared with controls (-12.9 +/- 3.6 and -15.4 +/- 5.1 vs -21.0 +/- 5.5 in transposition of the great arteries, congenitally corrected transposition of the great arteries, and controls, $P < .0001$). Systemic and pulmonary 2D-peak systolic strain were correlated in patients with transposition of the great arteries ($r = 0.46$, $P < .0001$) and congenitally corrected transposition of the great arteries ($r = 0.64$, $P < .0001$), suggesting interventricular interaction, and this was confirmed when EF on MRI was assessed ($r = 0.53$, $P < .0001$). More importantly, systemic 2D-peak systolic strain (hazard ratio 1.31, $P = .01$) was related to adverse clinical outcome (symptomatic progression to New York Heart Association class ≥ 3 , clinically relevant cardiac arrhythmia, or death) in patients with transposition of the great arteries and atrial switch independently of EF on CMR imaging, history of clinically relevant arrhythmia, or functional class.	3

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24. Ghelani SJ, Harrild DM, Gauvreau K, Geva T, Rathod RH. Comparison Between Echocardiography and Cardiac Magnetic Resonance Imaging in Predicting Transplant-Free Survival After the Fontan Operation. <i>Am J Cardiol.</i> 2015;116(7):1132-1138.	Observational-Dx	127 patients	To compare the echocardiographic and CMR parameters associated with death or transplantation in patients after the Fontan operation.	Of the 127 eligible patients, there were 12 end points (9%; 10 deaths and 2 listing for transplant). Median age was 16.8 years (interquartile range 12 to 23.1) with a median follow-up of 3.8 years (interquartile range 2.6 to 5.7). Among clinical parameters, protein-losing enteropathy had the strongest association with the outcome. Among echocardiographic variables, global circumferential strain showed the strongest association (hazard ratio 1.3 per unit change, 95% CI, 1.1 to 1.5, $P=0.001$, C-index 0.81), whereas among CMR variables indexed ventricular end-diastolic volume showed the strongest association with the outcome (hazard ratio 1.04 per 10 mL/BSA(1.3) increase in volume, 95% CI, 1.02 to 1.06, $P=0.001$, C-index 0.82). Cox proportional hazards analysis revealed echocardiography and CMR models to each individually have a higher predictive ability than the clinical model; however, in direct comparison, neither technique was superior.	3
25. Sreeram N, Sutherland GR, Geuskens R, et al. The role of transoesophageal echocardiography in adolescents and adults with congenital heart defects. <i>Eur Heart J.</i> 1991;12(2):231-240.	Review/Other-Dx	133 patients	To determine the potential role of TEE in the management of adolescents and adults with CHD.	TEE altered the diagnosis in 19 patients (14%) and provided additional information in 75 patients (56%) compared to transthoracic echocardiography.	4
26. Osawa K, Miyoshi T, Morimitsu Y, et al. Comprehensive assessment of morphology and severity of atrial septal defects in adults by CT. <i>J Cardiovasc Comput Tomogr.</i> 2015;9(4):354-361.	Observational-Dx	50 patients	To evaluate the usefulness of CT for assessing the hemodynamics of secundum ASD in adults compared with TEE, transthoracic echocardiography, and invasive catheterization.	The maximum sizes of the secundum ASDs derived from CT and TEE studies were comparable (21.2 +/- 8.0 vs 20.0 +/- 7.3 mm; $P=.41$; $r = 0.960$; $P<.001$). The rim lengths for the aortic, mitral, and tricuspid valves; the inferior vena cava; and posterior atrium were also comparable between CT and TEE measurements. The mean Qp/Qs ratio that was derived from CT measurements was comparable with that found by invasive catheterization (2.3 +/- 0.7 vs 2.3 +/- 0.8; $P=.73$; $r = 0.786$; $P<.001$).	3

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27. Flachskampf FA, Wouters PF, Edvardsen T, et al. Recommendations for transoesophageal echocardiography: EACVI update 2014. <i>Eur Heart J Cardiovasc Imaging</i> . 2014;15(4):353-365.	Review/Other-Dx	N/A	To update the recommendations for transoesophageal echocardiography of the European Association of Cardiovascular Imaging.	No results stated in abstract.	4
28. Daniel WG, Erbel R, Kasper W, et al. Safety of transesophageal echocardiography. A multicenter survey of 10,419 examinations. <i>Circulation</i> . 1991;83(3):817-821.	Observational-Tx	10,419 examinations at 15 centers	To analyze the experience of 15 European centers performing TEE studies for at least 1 year.	At the time of this survey, 10,419 TEE examinations had been attempted or performed in these institutions. These TEE examinations were carried out by 54 physicians, 53.7% of whom had been trained in endoscopic techniques. Within the same time period, 160,431 precordial echocardiographic examinations were performed in the 15 institutions; the ratio between TEE and transthoracic studies averaged 9.03 +/- 6.4% (range of the 15 centers, 1.4%–23.6%). Of the 10,419 patients, 9,240 (88.7%) were conscious inpatients or outpatients at the time of the TEE examination; the vast majority of the conscious patients did not receive IV sedation before TEE. In 201 cases (1.9%), insertion of the TEE probe was unsuccessfully attempted because of a lack of patient cooperation and/or operator experience (98.5%) or because of anatomical reasons (1.5%). In 90 of 10,218 TEE studies (0.88%) with successful probe insertion, the examination had to be interrupted because of the patient's intolerance of the echoscope (65 cases); because of pulmonary (8 cases), cardiac (8 cases), or bleeding complications (2 cases); or for other reasons (7 cases). 1 of the bleeding complications resulted from a malignant lung tumor with esophageal infiltration and was fatal (mortality rate, 0.0098%).	2
29. Esmaili A, Hohn R, Koch A, Vogl TJ, Hofstetter R, Abolmaali N. Assessment of shunt volumes in children with ventricular septal defects: comparative quantification of MR flow measurements and invasive oximetry. <i>Clin Res Cardiol</i> . 2006;95(10):523-530.	Observational-Dx	14 children	To compare invasive oximetry with MR based shunt volume quantification in pediatric patients of any age suffering from ventricular septal defects.	A good correlation between both methods was observed ($r(2) = 0.8$, $P < 0.0001$, CI 95%, = 0.62–1.22). A tendency towards higher values in the noninvasive technique was found in the Bland-Altman plot (bias = 3.79).	3

* See Last Page for Key

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30. Hundley WG, Li HF, Lange RA, et al. Assessment of left-to-right intracardiac shunting by velocity-encoded, phase-difference magnetic resonance imaging. A comparison with oximetric and indicator dilution techniques. <i>Circulation</i> . 1995;91(12):2955-2960.	Observational-Dx	21 subjects	To determine whether velocity-encoded, phase-difference MRI can assess the magnitude of intracardiac left-to-right shunting in humans.	Of the 21 patients, 12 had left-to-right intracardiac shunting detected by hydrogen inhalation. There was a good correlation ($r = .94$) between the invasive and MRI assessments of shunt magnitude. In comparison to oximetry and indocyanine green, MRI correctly identified the 12 patients with a ratio of pulmonary to systemic flow (Qp/Qs) of <1.5 (9 without intracardiac shunting and 3 with small shunts) and the 9 patients with a Qp/Qs of ≥ 1.5 (6 with ASD, 1 with ventricular septal defect, 1 with patent ductus arteriosus, and 1 with both ASD and patent ductus arteriosus).	1
31. Fratz S, Hess J, Schwaiger M, Martinoff S, Stern HC. More accurate quantification of pulmonary blood flow by magnetic resonance imaging than by lung perfusion scintigraphy in patients with fontan circulation. <i>Circulation</i> . 2002;106(12):1510-1513.	Observational-Dx	15 patients and 12 controls	To determine whether phase-velocity MRI is more accurate than scintigraphy for quantitative evaluation of pulmonary perfusion ratios in patients with atriopulmonary anastomosis or total cavopulmonary connection or partial cavopulmonary connection.	Bland-Altman analysis showed a clinically unacceptable difference of 7.1% right pulmonary blood flow (27.2% upper and -13.0% lower limit of agreement) between the 2 methods in the study group. The 2 methods agreed excellently in the control group (difference, 1.6%; 4.0% upper and -7.2% lower limit of agreement), showing that the bad agreement in the study group was caused by the problems encountered using pulmonary scintigraphy in patients with atriopulmonary anastomosis or total cavopulmonary connection or partial cavopulmonary connection.	3
32. Lubiszewska B, Gosiewska E, Hoffman P, et al. Myocardial perfusion and function of the systemic right ventricle in patients after atrial switch procedure for complete transposition: long-term follow-up. <i>J Am Coll Cardiol</i> . 2000;36(4):1365-1370.	Observational-Dx	61 patients	To assess RV function and identify patients with RV impairment long after Mustard or Senning operations for complete transposition of the great vessels using Tc-99m methoxyisobutyl isonitrile single-photon emission computed tomography.	Moderate to severe myocardial perfusion defects were noted in 20 patients at rest (33%) and an additional 13 patients (21%) with exercise. All patients demonstrated normal exercise tolerance. Myocardial perfusion defects correlated with impaired right and LV function. Larger perfusion defects were noted in older patients with longer follow-up. Authors suggest use of myocardial perfusion imaging to predict systemic ventricular impairment.	2

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33. Goo HW, Park IS, Ko JK, Kim YH, Seo DM, Park JJ. Computed tomography for the diagnosis of congenital heart disease in pediatric and adult patients. <i>Int J Cardiovasc Imaging</i> . 2005;21(2-3):347-365; discussion 367.	Review/Other-Dx	N/A	Review of MDCT experience for evaluation of CHD in children and adults.	MDCT can provide proper visualization of a variety of structural and functional abnormalities in patients with CHD.	4
34. Haramati LB, Glickstein JS, Issenberg HJ, Haramati N, Crooke GA. MR imaging and CT of vascular anomalies and connections in patients with congenital heart disease: significance in surgical planning. <i>Radiographics</i> . 2002;22(2):337-347; discussion 348-339.	Review/Other-Dx	23 patients	To illustrate the use of MRI and CT in the preoperative assessment of patients with CHD.	MRI and CT can provide helpful information for planning surgical repair in patients with CHD.	4
35. Memisoglu E, Hobikoglu G, Tepe MS, Norgaz T, Bilsel T. Congenital coronary anomalies in adults: comparison of anatomic course visualization by catheter angiography and electron beam CT. <i>Catheter Cardiovasc Interv</i> . 2005;66(1):34-42.	Observational-Dx	28 total patients: 14 with X-ray catheter angiography, 14 matched controls	To compare anatomic course of anomalous coronary arteries as seen on electron beam CTA and x-ray catheter angiography.	All normal and anomalous coronary artery origins were identified by both techniques. In 5 cases, there was discrepancy in the interpretation of the actual course of the coronary artery but consensus interpretation by cardiologists and radiologists favored the electron beam CTA interpretation over that of conventional angiography.	1
36. Ben Saad M, Rohnean A, Sigal-Cinqualbre A, Adler G, Paul JF. Evaluation of image quality and radiation dose of thoracic and coronary dual-source CT in 110 infants with congenital heart disease. <i>Pediatr Radiol</i> . 2009;39(7):668-676.	Observational-Dx	110 consecutive infants	To evaluate the image quality and radiation dose of thoracic and coronary dual-source CT in babies with CHD.	Mean dose-length product was 8+/-6 mGy x cm (effective dose 0.5+/-0.2 mSv) and 21+/-9 mGy x cm (effective dose 1.3+/-0.6 mSv) during the non-electrocardiography-gated spiral acquisition and electrocardiography-gated acquisition, respectively. Diagnostic quality images were achieved with the spiral acquisition in 89% of cases. Compared to the spiral mode, electrocardiography-gated acquisition significantly improved the visualization of the coronary arteries, with a diagnostic rate of 91% and 84% for the left and right coronary arteries, respectively. Dual-source CT together with iopromide at 300 mg/ml is a valuable tool for the routine clinical evaluation of infants with CHD. Electrocardiography-gated acquisition provides reliable visualization of the course of the coronary arteries.	3

**Known or Suspected Congenital Heart Disease in the Adult
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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
37. Leschka S, Oechslin E, Husmann L, et al. Pre- and postoperative evaluation of congenital heart disease in children and adults with 64-section CT. <i>Radiographics</i> . 2007;27(3):829-846.	Review/Other-Dx	N/A	To highlight the importance of knowledge about the anatomy, morphology, and terminology of CHD and to describe a sequential segmental approach for the accurate and comprehensive assessment of pediatric and adult patients with such disease.	The high spatial and temporal resolution provided by MDCT, combined with the short scanning times, enable the use of no sedation or only short-term sedation while evaluating patients with CHD. Consequently, CT has become useful for pre- and postsurgical evaluation of a wide variety of cardiac defects in pediatric and adult patients.	4
38. Spevak PJ, Johnson PT, Fishman EK. Surgically corrected congenital heart disease: utility of 64-MDCT. <i>AJR Am J Roentgenol</i> . 2008;191(3):854-861.	Review/Other-Dx	N/A	Review CT appearance of postoperative morphology and complications after surgical correction of congenital heart anomalies.	Echocardiography is typically the initial imaging technique used for CHD; however, some thoracic regions are beyond the imaging scope of echocardiography, particularly after surgical revision.	4
39. Han BK, Hlavacek AM, Kay WA, et al. Multi-institutional evaluation of the indications and radiation dose of functional cardiovascular computed tomography (CCT) imaging in congenital heart disease. <i>Int J Cardiovasc Imaging</i> . 2015:[E-pub ahead of print].	Review/Other-Dx	298 functional CTs	To compare patient and scanner characteristics, radiation dose estimates, and image quality of functional CT studies performed in CHD patients from 3 institutions (1/2007-3/2013).	The most common referral diagnosis were tetralogy of Fallot (33%), transposition complexes (24%) single ventricle heart disease (15%), and left sided obstruction (15%). The reason for cardiac CT was presence of pacemaker (60%), need for detailed coronary artery imaging (18%), metallic artifact in CMR (12%), evaluation of prosthetic valve function (4%), and claustrophobia or body mass index too large for the available MR scanner (6%). 266 (89.3%) scans allowed quantification of ventricular function, 25 (8.4%) scans allowed qualitative assessment of function, and 7 (2.3%) of the scans were nondiagnostic for functional analysis. Median dose length product was 399 mGy cm (186, 614), and median effective dose was 5.5 mSv (2.6, 8.5).	4

Known or Suspected Congenital Heart Disease in the Adult
EVIDENCE TABLE

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
40. Meinel FG, Henzler T, Schoepf UJ, et al. ECG-synchronized CT angiography in 324 consecutive pediatric patients: spectrum of indications and trends in radiation dose. <i>Pediatr Cardiol.</i> 2015;36(3):569-578.	Review/Other-Dx	324 pediatric patients	To describe the spectrum of indications for pediatric ECG-synchronized CTA, the main determinants of radiation exposure, and trends in radiation dose over time at a single, tertiary referral center.	The most common primary indications for the CTA examinations included known or suspected coronary pathologies (n = 166), complex CHD (n = 73), and aortic pathologies (n = 41). Median radiation exposure decreased from 12 mSv for patients examined in the years 2005-2007 to 1.2 mSv for patients examined in the years 2011-2013 ($P < 0.001$). Patients scanned using a tube potential of 80 kV (n = 259) had a significantly lower median radiation dose (1.4 mSv) compared to patients who were scanned at 100 kV (n = 46, median 6.3 mSv) or 120 kV (n = 19, median 19 mSv, $P < 0.001$). Tube voltage, followed by tube current and the method of ECG-synchronization were the strongest independent predictors of radiation dose.	4
41. Dillman JR, Ellis JH, Cohan RH, Strouse PJ, Jan SC. Frequency and severity of acute allergic-like reactions to gadolinium-containing i.v. contrast media in children and adults. <i>AJR Am J Roentgenol.</i> 2007;189(6):1533-1538.	Observational-Tx	78,353 IV (65,009 adult and 13,344 pediatric) IV administrations of gadolinium-containing contrast material	Retrospective study to determine the frequency and severity of acute allergic-like reactions to IV-administered gadolinium-containing contrast media in children and adults.	48 reactions involved adult patients (adult reaction frequency, 0.07%), and 6 reactions occurred in pediatric patients (pediatric reaction frequency, 0.04%). 40 (74%) acute allergic-like reactions were mild, 10 (19%) were moderate, and 4 (7%) were severe. Adult and pediatric acute allergic-like reactions to IV-administered gadolinium-containing contrast media are rare. Most of these reactions are mild; however, moderate and severe reactions that require immediate management do occur.	3

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EVIDENCE TABLE

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
42. Nguyen SA, Suranyi P, Ravenel JG, et al. Iso-osmolality versus low-osmolality iodinated contrast medium at intravenous contrast-enhanced CT: effect on kidney function. <i>Radiology</i> . 2008;248(1):97-105.	Experimental-Tx	117 patients had contrast-enhanced CT with either iso-osmolality iodixanol (n=61) or low-osmolality iopromide (n=56)	Single-center, randomized, double-blind prospective study to investigate the effects of iso-osmolality contrast medium compared with a low-osmolality agent on renal function (SCr and glomerular filtration rate) in high-risk patients undergoing IV contrast-enhanced CT.	Iodixanol decreased SCr (mean +/- standard deviation) from 1.77 mg/dL +/- 0.24 (156.47 micromol/L +/- 21.22) at baseline to 1.65 mg/dL +/- 0.35 (145.86 micromol/L +/- 30.94, $P=.046$) at day 1, 1.73 mg/dL +/- 0.53 (152.93 micromol/L +/- 46.85, not significant) at day 2, and 1.73 mg/dL +/- 0.55 (152.93 micromol/L +/- 48.62, not significant) at day 3 (not significant). Iopromide increased SCr from 1.75 mg/dL +/- 0.32 (154.7 micromol/L +/- 28.29) at baseline to 1.8 mg/dL +/- 0.42 (159.12 micromol/L +/- 15.59) at day 1, 1.77 mg/dL +/- 0.49 (156.47 micromol/L +/- 43.32) at day 2, and 1.77 mg/dL +/- 0.62 (156.47 micromol/L +/- 54.81) at day 3 (not significant). Iodixanol increased and iopromide decreased glomerular filtration rate on all 3 days after CT (not significant). Fewer patients in the iodixanol group (8.5%) than in the iopromide group (27.8%) had SCr increase 0.5 mg/dL or higher ($\geq 25\%$, $P=.012$). 2 patients in each group had SCr increase of 1.0 mg/dL or more (not significant). More patients in the iopromide group (42.3%) than in the iodixanol group (24.1%) had a glomerular filtration rate reduction of 5 mL/min or higher ($P=.0426$). No patient had a contrast material-related adverse event at 30- or 90-day follow-up. IV contrast material application in high-risk patients is unlikely to be associated with permanent adverse outcomes. SCr levels after contrast material administration are lower in iodixanol than iopromide groups.	1
43. Haramati LB, Moche IE, Rivera VT, et al. Computed tomography of partial anomalous pulmonary venous connection in adults. <i>J Comput Assist Tomogr</i> . 2003;27(5):743-749.	Observational-Dx	29 cases from 1,825 chest CT reports	To determine incidence of partial anomalous venous return seen on 1,825 consecutive CT scans in adults (July 2000–July 2001).	A partial anomalous venous connection was seen in 0.2% of adults undergoing chest CT and most commonly left upper lobe (80%), female (66%) and infrequently associated with ASD (1%).	3

Known or Suspected Congenital Heart Disease in the Adult
EVIDENCE TABLE

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
44. Schmitt R, Froehner S, Brunn J, et al. Congenital anomalies of the coronary arteries: imaging with contrast-enhanced, multidetector computed tomography. <i>Eur Radiol.</i> 2005;15(6):1110-1121.	Observational-Dx	1,758: 44 selected from total	To determine ability of MDCT to detect and characterize anomalous coronary arteries.	44 anomalies (2.5%) of the coronary arteries were identified from 1,758 patient studies. 13 patients (0.74%) had “malignant” interartirial anomalous coronary arteries. Of note, selective cannulation was successful in only 11/20 (55%) of patients who underwent cardiac catheterization for final diagnosis.	3
45. Cook SC, Dyke PC, 2nd, Raman SV. Management of adults with congenital heart disease with cardiovascular computed tomography. <i>J Cardiovasc Comput Tomogr.</i> 2008;2(1):12-22.	Review/Other-Dx	87 patients	To evaluate the role of cardiac CT in the care of ACHD.	54 subjects had cardiac CT findings that warranted possible transcatheter or surgical interventions or both. Of these, 30 patients went on to transcatheter or surgical intervention based on cardiac CT results. 20 (37%) subjects of the study population had either pacemaker or implantable cardioverter defibrillator. Cardiac CT provides important volumetric imaging information to guide management in adult patients with CHD.	4
46. Hayabuchi Y, Inoue M, Watanabe N, et al. Assessment of systemic-pulmonary collateral arteries in children with cyanotic congenital heart disease using multidetector-row computed tomography: comparison with conventional angiography. <i>Int J Cardiol.</i> 2010;138(3):266-271.	Observational-Dx	48 consecutive patients	To assess the feasibility of MDCT for the evaluation of systemic-pulmonary collateral arteries in children with CHD associated with reduced pulmonary blood flow. Gold standard used was conventional angiographic findings. MDCT evaluated by 2 reviewers.	115 systemic-pulmonary collateral arteries were identified with conventional angiography, and 94 systemic-pulmonary collateral arteries were identified with MDCT. In 89 (77%) vessels, concordant findings were observed with both modalities, with adequate depiction in 53 vessels and suboptimal depiction in 36 vessels. In 26 (23%) vessels, MDCT was unable to identify systemic-pulmonary collateral arteries. Further, CTA resulted in the false-positive identification of vessels in 5 cases. Excellent correlation between MDCT- and conventional angiography-based measurement of systemic-pulmonary collateral vessel diameter ($R(2)=0.83$), although a systematic overestimation was observed with MDCT (bias 0.19+/-0.74 mm). MDCT is a potentially useful tool, which may have implications for planning percutaneous interventions and surgical repair in the future.	2

Known or Suspected Congenital Heart Disease in the Adult
EVIDENCE TABLE

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
47. Hlavacek AM. Imaging of congenital cardiovascular disease: the case for computed tomography. <i>J Thorac Imaging</i> . 2010;25(3):247-255.	Review/Other-Dx	N/A	To review the utility of CTA and address the risks and benefits of this modality in comparison with other imaging modalities.	CTA provides important advantages over echocardiography and MRI modalities in certain clinical scenarios. CTA is a useful adjunct to echocardiography in patients with CHD and is often preferable to MRI in certain patients.	4
48. Garg R, Powell AJ, Sena L, Marshall AC, Geva T. Effects of metallic implants on magnetic resonance imaging evaluation of Fontan palliation. <i>Am J Cardiol</i> . 2005;95(5):688-691.	Observational-Dx	101 consecutive patients	To examine the effects of endovascular metallic implants on the ability of MRI to evaluate cardiovascular anatomy and to assess ventricular size and function in patients with Fontan palliation.	Compared with studies in patients without implants, in whom 100% of anatomic segments were adequately visualized and the median image quality grade for ventricular function assessment was 5 of 5, the presence of any implant (54% of studies) reduced the proportion of adequately imaged anatomy to 77% ($P<.001$) and the median image quality grade for ventricular function assessment to 3 of 5 ($P<.001$).	3
49. Felmeden D, Singh SP, Lip GY. Anomalous coronary arteries of aortic origin. <i>Int J Clin Pract</i> . 2000;54(6):390-394.	Review/Other-Dx	N/A	To review anomalous coronary arteries of aortic origin.	It is important to identify the exact cardiac anatomy, particularly in patients undergoing angioplasty, stenting or cardiac surgery.	4
50. Xu H, Zhu Y, Zhu X, Tang L, Xu Y. Anomalous coronary arteries: depiction at dual-source computed tomographic coronary angiography. <i>J Thorac Cardiovasc Surg</i> . 2012;143(6):1286-1291.	Observational-Dx	124 patients	To retrospectively determine the imaging features of coronary artery anomalies depicted at dual-source CCTA.	There were 124 (1.02%) patients with coronary anomalies. 51 patients demonstrated an anomalous origin of the right coronary artery from the left sinus of Valsalva or the left main artery. An anomalous origin of a left circumflex artery from the right sinus of Valsalva or the right coronary artery was depicted in 17 patients. An anomalous origin of a left main artery from the right sinus of Valsalva was depicted in 1 patient. A single coronary artery was shown in 4 patients, and congenital transposition of the great arteries was associated with this anomaly in 1 patient. In the remaining 50 patients, coronary artery fistulas were identified. 8 patients were referred after an equivocal conventional coronary angiogram.	4

Known or Suspected Congenital Heart Disease in the Adult
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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
51. Szymczyk K, Polgaj M, Szymczyk E, Majos A, Grzelak P, Stefanczyk L. Prevalence of congenital coronary artery anomalies and variants in 726 consecutive patients based on 64-slice coronary computed tomography angiography. <i>Folia Morphol (Warsz)</i> . 2014;73(1):51-57.	Review/Other-Dx	726 patients	To determine the prevalence of coronary artery variations detected by 64-slice MDCT.	The overall incidence of coronary artery anomalies was 1.1% (8 out of 726 participants). The most common anomaly was an anomalous origin of the circumflex artery from the right coronary sinus with a retroaortic course (4 patients, 0.6%), followed by origin of right coronary artery from the left coronary sinus (2 patients, 0.3%). 1 patient with abnormal origin of the left main artery from the right coronary sinus (0.1%) and 1 patient with a circumflex artery origin from the proximal segment of the right coronary artery (0.1%) were observed, both with retroaortic course.	4
52. Zenooz NA, Habibi R, Mammen L, Finn JP, Gilkeson RC. Coronary artery fistulas: CT findings. <i>Radiographics</i> . 2009;29(3):781-789.	Review/Other-Dx	N/A	To present the pathophysiology, clinical features, imaging diagnosis, and treatment of different types of coronary artery fistulas, with emphasis on common anatomic sites and cross-sectional imaging features.	Although conventional angiography has been used for evaluation of the coronary arteries, cross-sectional imaging is a noninvasive and useful modality for diagnosis of coronary artery fistulas. Therefore, in addition to evaluation of the coronary arteries for stenosis and plaques, special attention should be paid to their courses and terminations in every CT study of the heart to detect these potentially fatal anomalies.	4
53. Meijboom WB, van Mieghem CA, Mollet NR, et al. 64-slice computed tomography coronary angiography in patients with high, intermediate, or low pretest probability of significant coronary artery disease. <i>J Am Coll Cardiol</i> . 2007;50(15):1469-1475.	Observational-Dx	254 patients	To assess the usefulness of 64-slice CCTA to detect or rule out CAD in patients with various estimated pretest probabilities of CAD.	The estimated pretest probability of CAD in the high (n = 105), intermediate (n = 83), and low (n = 66) groups was 87%, 53%, and 13%, respectively. The diagnostic performance of the CT scan was different in the 3 subgroups. The estimated post-test probability of the presence of significant CAD after a negative CT scan was 17%, 0%, and 0% and after a positive CT scan was 96%, 88%, and 68%, respectively.	3

**Known or Suspected Congenital Heart Disease in the Adult
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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
54. Budoff MJ, Dowe D, Jollis JG, et al. Diagnostic performance of 64-multidetector row coronary computed tomographic angiography for evaluation of coronary artery stenosis in individuals without known coronary artery disease: results from the prospective multicenter ACCURACY (Assessment by Coronary Computed Tomographic Angiography of Individuals Undergoing Invasive Coronary Angiography) trial. <i>J Am Coll Cardiol.</i> 2008;52(21):1724-1732.	Experimental-Dx	230 patients	To evaluate the diagnostic accuracy of electrocardiographically gated CCTA in individuals without known CAD.	On a patient-based model, the sensitivity, specificity, and positive and negative predictive values to detect $\geq 50\%$ or $\geq 70\%$ stenosis were 95%, 83%, 64%, and 99%, respectively, and 94%, 83%, 48%, 99%, respectively. No differences in sensitivity and specificity were noted for nonobese compared with obese subjects or for heart rates ≤ 65 beats/min compared with >65 beats/min, whereas calcium scores >400 reduced specificity significantly.	1
55. Douglas PS, Hoffmann U, Patel MR, et al. Outcomes of anatomical versus functional testing for coronary artery disease. <i>N Engl J Med.</i> 2015;372(14):1291-1300.	Experimental-Dx	10,003 patients	The objective of the Prospective Multicenter Imaging Study for Evaluation of Chest Pain (PROMISE) was to compare health outcomes in patients who presented with new symptoms suggestive of CAD that required further evaluation and who was randomly assigned to an initial strategy of anatomical testing with the use of CTA or to functional testing.	The mean age of the patients was 60.8 \pm 8.3 years, 52.7% were women, and 87.7% had chest pain or dyspnea on exertion. The mean pretest likelihood of obstructive CAD was 53.3 \pm 21.4%. Over a median follow-up period of 25 months, a primary end-point event occurred in 164/4996 patients in the CTA group (3.3%) and in 151/5007 (3.0%) in the functional-testing group (adjusted hazard ratio, 1.04; 95% CI, 0.83 to 1.29; $P=0.75$). CTA was associated with fewer catheterizations showing no obstructive CAD than was functional testing (3.4% vs 4.3%, $P=0.02$), although more patients in the CTA group underwent catheterization within 90 days after randomization (12.2% vs 8.1%). The median cumulative radiation exposure per patient was lower in the CTA group than in the functional-testing group (10.0 mSv vs 11.3 mSv), but 32.6% of the patients in the functional-testing group had no exposure, so the overall exposure was higher in the CTA group (mean, 12.0 mSv vs 10.1 mSv; $P<0.001$).	1

Known or Suspected Congenital Heart Disease in the Adult
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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
56. Gerber TC, Kantor B, McCollough CH. Radiation dose and safety in cardiac computed tomography. <i>Cardiol Clin.</i> 2009;27(4):665-677.	Review/Other-Dx	N/A	To review the existing data regarding biologic hazards of radiation exposure associated with medical diagnostic testing, the methodologies used to estimate radiation exposure and dose, and the measures that can be taken to effectively reduce that exposure.	The risk of causing a malignancy at the radiation dose levels used in cardiac imaging is hypothetical, not proven, and estimates of radiation dose have a wide margin of error. However, in the absence of certainty, the consensus opinions of influential expert panels advocate adopting a conservative estimate of radiation risks.	4
57. Earls JP, Berman EL, Urban BA, et al. Prospectively gated transverse coronary CT angiography versus retrospectively gated helical technique: improved image quality and reduced radiation dose. <i>Radiology.</i> 2008;246(3):742-753.	Observational-Dx	203 patients	To retrospectively compare image quality, radiation dose, and blood vessel assessability for CCTA obtained with a prospectively gated transverse CT technique and a retrospectively gated helical CT technique.	The mean effective dose for the group with the prospectively gated transverse technique was 2.8 mSv; this represents an 83% reduction as compared with that for the group with the retrospectively gated helical technique (mean, 18.4 mSv; $P < .001$). The image quality score for each of the arteries, as well as the overall combined score, was significantly greater for images obtained with prospectively gated transverse technique than for images obtained with retrospectively gated helical technique. The combined mean image quality score was 4.791 for images obtained with prospectively gated transverse technique vs 4.514 for images obtained with retrospectively gated helical technique (proportional odds model OR, 2.8; 95% CI: 1.7, 4.8). The percentage of assessable coronary artery segments was 98.6% (1,196/1,213) for images obtained with prospectively gated transverse technique vs 97.9% (1,741/1,778) for images obtained with retrospectively gated helical technique ($P = .83$).	2

Known or Suspected Congenital Heart Disease in the Adult
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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
58. Husmann L, Valenta I, Gaemperli O, et al. Feasibility of low-dose coronary CT angiography: first experience with prospective ECG-gating. <i>Eur Heart J</i> . 2008;29(2):191-197.	Observational-Dx	41 consecutive patients	To determine the feasibility of prospective ECG-gating to achieve low-dose CCTA.	Mean effective radiation dose was 2.1 +/- 0.6 mSv (range, 1.1-3.0 mSv). Image quality was inversely related to heart rate (57.3 +/- 6.2, range 39-66 b.p.m.; $r = 0.58$, $P < 0.001$), vessel attenuation (346 +/- 104, range 110-780 HU; $r = 0.56$, $P < 0.001$), and body mass index (26.1 +/- 4.0, range 19.1-36.3 kg/m ²); $r = 0.45$, $P < 0.001$), but not to heart rate variability (1.5 +/- 1.0, range 0.2-5.1 b.p.m.; $r = 0.28$, $P = 0.069$). Non-diagnostic CCTA image quality was found in 5.0% of coronary segments. However, below a heart rate of 63 b.p.m. (n=28), as determined ROC, only 1.1% of coronary segments were non-diagnostic compared with 14.8% with heart rate of >63 b.p.m. ($P < 0.001$).	3
59. Stolzmann P, Leschka S, Scheffel H, et al. Dual-source CT in step-and-shoot mode: noninvasive coronary angiography with low radiation dose. <i>Radiology</i> . 2008;249(1):71-80.	Observational-Dx	40 patients	To prospectively investigate CT image quality parameters by using different protocols and to calculate radiation dose estimates for noninvasive coronary angiography performed with dual-source CT in the step-and-shoot mode.	Mean image noise was similar with protocols A and B. Mean attenuation in the aorta and coronary arteries with protocol A (444 HU) was significantly ($P < .001$) higher than that with protocol B (358 HU). The reduced contrast material dose in protocol C yielded attenuation similar to that with protocol B. Diagnostic image quality was achieved with all protocols in 1,237 (97.9%) of 1,264 coronary segments. No significant differences in image quality between the 100- and 120-kV protocols were found. Mean heart rate had a significant effect on motion artifacts (AUC = 0.818; 95% CI: 0.723, 0.892; $P < .001$), whereas heart rate variability had a significant effect on stair-step artifacts (AUC = 0.79; 95% CI: 0.687, 0.865; $P < .001$). The mean estimated effective dose was 1.2 mSv +/- 0.2 for protocols A and C and 2.6 mSv +/- 0.5 for protocol B.	2

**Known or Suspected Congenital Heart Disease in the Adult
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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
60. Leipsic J, Labounty TM, Heilbron B, et al. Estimated radiation dose reduction using adaptive statistical iterative reconstruction in coronary CT angiography: the ERASIR study. <i>AJR Am J Roentgenol.</i> 2010;195(3):655-660.	Observational-Dx	574 consecutive patients	Prospectively evaluate patients undergoing CCTA at 3 centers to assess the impact of Adaptive Statistical Iterative Reconstruction (ASIR) on radiation dose and study quality for CCTA. Comparisons were performed between consecutive groups initially using filtered back projection (n = 331) and subsequently ASIR (n = 243) with regard to patient and scan characteristics, radiation dose, and diagnostic study quality.	There was no difference between groups in the use of prospective gating, tube voltage, or scan length. The examinations performed using ASIR had a lower median tube current than those obtained using filtered back projection (median [interquartile range], 450 mA [350–600] vs 650 mA [531–750], respectively; $P < 0.001$). There was a 44% reduction in the median radiation dose between the filtered back projection and ASIR cohorts (4.1 mSv [2.3–5.2] vs 2.3 mSv [1.9–3.5]; $P < 0.001$). After adjustment for scan settings, ASIR was associated with a 27% reduction in radiation dose compared with filtered back projection (95% CI, 21%–32%; $P < 0.001$). Despite the reduced current, ASIR was not associated with a difference in adjusted signal, noise, or signal-to-noise ratio ($P = \text{not significant}$). No differences existed between filtered back projection and ASIR for interpretability per coronary artery (98.5% vs 99.3%, respectively; $P = 0.12$) or per patient (96.1% vs 97.1%, $P = 0.65$). ASIR enabled reduced tube current and lower radiation dose in comparison with filtered back projection, with preserved signal, noise, and study interpretability, in a large multicenter cohort. ASIR represents a new technique to reduce radiation dose in CCTA studies.	3
61. Achenbach S, Marwan M, Ropers D, et al. Coronary computed tomography angiography with a consistent dose below 1 mSv using prospectively electrocardiogram-triggered high-pitch spiral acquisition. <i>Eur Heart J.</i> 2010;31(3):340-346.	Observational-Dx	50 consecutive patients	To evaluate the feasibility and image quality of a new scan mode for CCTA with an effective dose of < 1 mSv.	In all 50 patients, imaging was successful. Mean duration of data acquisition was 258 +/- 20 ms. Mean dose-length product was 62 +/- 5 mGy cm, the effective dose was 0.87 +/- 0.07 mSv (0.78-0.99 mSv). Of the 742 coronary artery segments, 94% had an image quality score of 1, 5.0% a score of 2, 0.9% a score of 3, and 4 segments (0.5%) were 'uninterpretable'. In non-obese patients with a low and stable heart rate, prospectively ECG-triggered high-pitch spiral CCTA provides excellent image quality at a consistent dose below 1.0 mSv.	3

Known or Suspected Congenital Heart Disease in the Adult
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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
62. Beerbaum P, Korperich H, Gieseke J, Barth P, Peuster M, Meyer H. Rapid left-to-right shunt quantification in children by phase-contrast magnetic resonance imaging combined with sensitivity encoding (SENSE). <i>Circulation</i> . 2003;108(11):1355-1361.	Observational-Dx	22 pediatric patients	To determine feasibility of using parallel imaging for acceleration of phase contrast MR measurement of left-to-right shunts (Qp/Qs ratio).	Phase contrast MR measurements using parallel imaging accelerated acquisitions with negligible differences ($\pm 3\%$) in Qp/Qs ratios vs standard phase contrast MR without parallel imaging.	3
63. Dodge-Khatami A, Tulevski, II, Bennink GB, et al. Comparable systemic ventricular function in healthy adults and patients with unoperated congenitally corrected transposition using MRI dobutamine stress testing. <i>Ann Thorac Surg</i> . 2002;73(6):1759-1764.	Observational-Dx	24 patients, 11 controls	To determine systemic RV function using dobutamine stress in patients with congenitally corrected transposition of the great vessels (n=13) vs healthy controls.	Compared to healthy controls, patients with congenitally corrected transposition of the great vessels had larger systemic RV volumes, diminished EFs but appropriate response to dobutamine.	3
64. Dyme JL, Prakash A, Printz BF, Kaur A, Parness IA, Nielsen JC. Physiology of isolated anomalous pulmonary venous connection of a single pulmonary vein as determined by cardiac magnetic resonance imaging. <i>Am J Cardiol</i> . 2006;98(1):107-110.	Observational-Dx	6 patients	To determine physiological effects of isolated anomalous pulmonary venous return using MR.	Patients with isolated anomalous pulmonary venous return were found to have modest left-to-right shunt (Qp/Qs ratio, 1.3 to 1.6) with mild dilation of the RV.	3
65. Fogel MA, Hubbard A, Weinberg PM. A simplified approach for assessment of intracardiac baffles and extracardiac conduits in congenital heart surgery with two- and three-dimensional magnetic resonance imaging. <i>Am Heart J</i> . 2001;142(6):1028-1036.	Review/Other-Dx	139 patients	To describe experience using MRI to evaluate intracardiac baffles and extracardiac conduits in patients with CHD.	From black blood spin echo and bright blood cine data, all 116 baffles and 28 conduits were successfully evaluated with assistance of multi-planar reconstruction and 3D reconstruction tools.	4
66. Geva T, Greil GF, Marshall AC, Landzberg M, Powell AJ. Gadolinium-enhanced 3-dimensional magnetic resonance angiography of pulmonary blood supply in patients with complex pulmonary stenosis or atresia: comparison with x-ray angiography. <i>Circulation</i> . 2002;106(4):473-478.	Observational-Dx	32 patients	To determine whether Gd-enhanced 3D MRA can provide noninvasive alternative to catheter x-ray angiography for evaluation of pulmonary arteries and APCs.	Compared with catheter angiography and surgical observations, Gd-enhanced 3D MRA was found to have a 100% sensitivity and 100% specificity for diagnosis of main (n=10) and branch vessel pulmonary artery stenosis or hypoplasia (n=38), and absent (n=5) or discontinuous (n=4) branch pulmonary arteries. All 48 APCs diagnosed at catheterization were diagnosed by MRA; MRA revealed 3 additional APCs not seen at catheterization. The mean difference in pulmonary vessel diameter measurement between MRA and catheterization was 0.5 ± 1.5 mm (mean interobserver difference = 0.4 ± 1.5 mm).	2

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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
67. Greil GF, Powell AJ, Gildein HP, Geva T. Gadolinium-enhanced three-dimensional magnetic resonance angiography of pulmonary and systemic venous anomalies. <i>J Am Coll Cardiol</i> . 2002;39(2):335-341.	Observational-Dx	61 patients	To determine diagnostic value of Gd-enhanced 3D MRA in patients with congenital and acquired anomalies of the pulmonary and systemic veins.	Compared to all available diagnostic data by other modalities, Gd-enhanced MRA detected all known or suspected pulmonary and systemic venous anomalies and provided new or additional information in 74% of patients. In 3 patients, catheterization did not detect venous anomalies subsequently identified on 3D MRA. In 17 patients (28%), unsuspected diagnosis of a venous anomaly was identified on MRA. In another 28 patients (46%), suspected diagnosis was confirmed and additional clinically important information was provided on MRA.	3
68. Mohrs OK, Petersen SE, Voigtlaender T, et al. Time-resolved contrast-enhanced MR angiography of the thorax in adults with congenital heart disease. <i>AJR Am J Roentgenol</i> . 2006;187(4):1107-1114.	Observational-Dx	20 patients	To evaluate the diagnostic value of a breath hold time-resolved Gd-enhanced MRA technique in the evaluation of ACHD.	Time-resolved MRA was found to provide a very high diagnostic value (92% of diagnostic parameters assessed) that included thoracic vascular anatomy, sequential cardiac anatomy, and shunt detection. For all 3 categories, high sensitivity (93%–100%) and specificity (87%–100%) was found.	2
69. Oosterhof T, Tulevski, II, Roest AA, et al. Disparity between dobutamine stress and physical exercise magnetic resonance imaging in patients with an intra-atrial correction for transposition of the great arteries. <i>J Cardiovasc Magn Reson</i> . 2005;7(2):383-389.	Experimental-Dx	64 total patients: 39 cohort, 25 healthy controls	To compare the hemodynamic response of dobutamine and submaximal exercise MRI in patients with intra-atrial correction of transposition of the great vessels.	In the healthy cohort, the dobutamine and exercise MR studies showed similar responses. However, in the patient cohort, there were significant differences in response (EF, end-diastolic volume, end-systolic volume, wall thickening) were noted between dobutamine stress and exercise stress MRI; such that the authors suggest that the results of the 2 types of stress are not necessarily interchangeable in the clinical setting.	2
70. Prasad SK, Soukias N, Hornung T, et al. Role of magnetic resonance angiography in the diagnosis of major aortopulmonary collateral arteries and partial anomalous pulmonary venous drainage. <i>Circulation</i> . 2004;109(2):207-214.	Observational-Dx	29 patients	To determine major APCs in patients with CHD using Gd-enhanced 3D MRA.	All APCs were correctly identified on Gd-enhanced 3D MRA. Compared to cardiac catheterization, echocardiogram and surgical inspection, Gd-enhanced 3D MRA provided additional information related to confluence and size of the pulmonary arteries (n=13), pulmonary artery stenosis (n=3), aneurismal dilatation of the pulmonary artery (n=1), and additional anomalous vascular abnormality (n=3).	2

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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
71. Haggerty CM, de Zelicourt DA, Restrepo M, et al. Comparing pre- and post-operative Fontan hemodynamic simulations: implications for the reliability of surgical planning. <i>Ann Biomed Eng.</i> 2012;40(12):2639-2651.	Experimental-Tx	4 patients	To quantify the impact of the numerous unknown variables inherent to prospective surgical modeling (eg, geometric agreement, adaptation of input and output boundary conditions) to assess the predictive power of the virtual surgery paradigm.	Despite variations in physiologic boundary conditions (eg, cardiac output, venous flows) and the exact geometry of the surgical baffle, sufficient agreement was observed with respect to hepatic flow distribution (90% CI, -14 +/- 4.3% difference). There was also good agreement of flow-normalized energetic efficiency predictions (19 +/- 4.8% error).	2
72. Lee VS, Resnick D, Bundy JM, Simonetti OP, Lee P, Weinreb JC. Cardiac function: MR evaluation in one breath hold with real-time true fast imaging with steady-state precession. <i>Radiology.</i> 2002;222(3):835-842.	Observational-Dx	20 total: 8 cohort, 12 healthy controls	To compare real-time and single-slice fast imaging with steady state free precession to traditional single-slice fast gradient echo imaging for cardiac assessment.	Myocardial-to-blood contrast-to-noise ratios were superior for both steady state free precession techniques vs traditional fast gradient echo imaging. Resting LV myocardial function with real-time steady state free precession was comparable to that measured using single-slice steady state free precession.	3
73. Lyen S, Mathias H, McAlindon E, et al. Optimising the imaging plane for right ventricular magnetic resonance volume analysis in adult patients referred for assessment of right ventricular structure and function. <i>J Med Imaging Radiat Oncol.</i> 2015;59(4):421-430.	Observational-Dx	20 patients	To evaluate the reproducibility and accuracy of using short-axis and axial (transaxial) plane for MRI analysis in adult patients referred for assessment of RV structure and function.	The concordance between the RV stroke volumes and LV stroke volumes was good using both methods (axial RV stroke volumes concordance correlation coefficients = 0.93, short-axis RV stroke volumes concordance correlation coefficients = 0.86). Paired t-test and analysis of variance showed that the LV/RV stroke volume differences were not significant ($P=0.17$). There was slight improvement in interobserver reliability with end systolic volume measurements (axial intraclass correlation coefficients = 0.92, short-axis intraclass correlation coefficients = 0.81) but this failed to reach statistical significance ($P=0.37$). There was excellent intraobserver variability (intraclass correlation coefficients >0.9).	2
74. Moody WE, Edwards NC, Chue CD, et al. Variability in cardiac MR measurement of left ventricular ejection fraction, volumes and mass in healthy adults: defining a significant change at 1 year. <i>Br J Radiol.</i> 2015;88(1049):20140831.	Observational-Dx	42 healthy subjects	To examine the within-subject changes in LV volumes, LV mass and EF using CMR over 12 months in a well-characterized cohort of healthy adults.	There were no significant changes in any LV parameter on repeat CMR at 12 months. The short-term interstudy biases were not significantly different from the long-term changes observed at 1 year. The smallest detectable change for LVEF, end-diastolic volume, end-systolic volume and LVM that could be recognized with 95% confidence were 6%, 13 mL, 7 mL and 6-g, respectively.	3

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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
75. Makowski MR, Wiethoff AJ, Uribe S, et al. Congenital heart disease: cardiovascular MR imaging by using an intravascular blood pool contrast agent. <i>Radiology</i> . 2011;260(3):680-688.	Observational-Dx	23 patients	To compare the image quality and diagnostic performance of a contrast agent-specific inversion-recovery steady-state free precession MRI sequence performed by using an intravascular contrast agent (gadofosveset trisodium) with those of a commonly used T2-prepared steady-state free precession sequence performed by using an extravascular (gadopentetate dimeglumine) and an intravascular (gadofosveset trisodium) contrast agent in patients with CHD.	Use of the inversion-recovery steady-state free precession sequence with gadofosveset trisodium significantly improved vessel wall sharpness, contrast to noise ratios, and image quality ($P < .05$ for all) for all investigated intra- and extracardiac structures compared with the T2-prepared steady-state free precession sequence with gadopentetate dimeglumine and gadofosveset trisodium and the respective contrast-enhanced MR angiographic examinations. With use of the inversion-recovery steady-state free precession sequence with gadofosveset trisodium, new, unsuspected diseases (5 [22%] of 23) were diagnosed, while other diseases could be excluded (15 [65%] of 23). Information available from echocardiography (n = 23), conventional angiography (n = 4), and/or surgery (n = 1) confirmed all diagnoses.	3
76. Naehle CP, Kaestner M, Muller A, et al. First-pass and steady-state MR angiography of thoracic vasculature in children and adolescents. <i>JACC Cardiovasc Imaging</i> . 2010;3(5):504-513.	Observational-Dx	25 patients	To compare a high-resolution, ECG-gated, motion-compensated, steady-state-MRA of the thoracic vasculature to standard first-pass-MRA using a blood-pool contrast agent (gadofosveset trisodium).	Steady-state-MRA yielded significantly higher image quality for all vessels, and significantly higher vessel sharpness for LSPV and LPA compared to the first-pass-MRA. Steady-state-MRA revealed lower intra- and interobserver variability for vessel diameters compared with the first-pass-MRA. The first-pass-MRA showed higher CR compared to the steady-state-MRA.	2
77. Bunce NH, Lorenz CH, Keegan J, et al. Coronary artery anomalies: assessment with free-breathing three-dimensional coronary MR angiography. <i>Radiology</i> . 2003;227(1):201-208.	Observational-Dx	26 patients	To evaluate a simplified protocol by using free-breathing 3D coronary MRA to determine the anatomy of anomalous coronary arteries, in particular the relationship of the vessels to the aortic root.	6 anomalous circumflex arteries originated from the right sinus of Valsalva and passed behind the aortic root. 6 right coronary arteries arose from the left sinus of Valsalva and coursed between the aortic root and the RV outflow tract. 9 left coronary arteries arose from the right sinus of Valsalva; 7 of 9 coursed between the aortic root and the RV outflow tract. 5 patients had minor anomalies. Overall, in 8 patients with anomalous arteries that coursed between the aortic root and the RV outflow tract, conventional coronary angiography could not be used confidently to identify the proximal course.	3

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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
78. Glatz AC, Rome JJ, Small AJ, et al. Systemic-to-pulmonary collateral flow, as measured by cardiac magnetic resonance imaging, is associated with acute post-Fontan clinical outcomes. <i>Circ Cardiovasc Imaging</i> . 2012;5(2):218-225.	Observational-Dx	44 subjects	To evaluate the association between systemic-to-pulmonary flow and acute post-Fontan clinical outcomes using a novel method of quantifying systemic-pulmonary collateral flow by CMR imaging.	Systemic-pulmonary collateral flow prior to Fontan measured 1.5+/-0.9 L/min/m(2), accounting for 31+/-11% of total aortic flow and 44+/-15% of total pulmonary venous flow. There was a significant linear association between natural log-transformed duration of hospitalization and systemic-pulmonary collateral flow as a proportion of total aortic (rho=0.31, P=0.04) and total pulmonary venous flow (rho=0.29, P=0.05). After adjustment for Fontan type and presence of a fenestration, absolute systemic-pulmonary collateral flow was significantly associated with hospital duration ≥7 days (OR=9.2, P=0.02) and chest tube duration ≥10 days (OR=22.7, P=0.009). Similar associations exist for systemic-pulmonary collateral flow as a percentage of total aortic (OR=1.09, P=0.048 for hospitalization ≥7 days; OR=1.24, P=0.007 for chest tube duration ≥10 days) and total pulmonary venous flow (OR=1.07, P=0.048 for hospitalization ≥7 days; OR=1.18, P=0.006 for chest tube duration ≥10 days).	3
79. Bell A, Beerbaum P, Greil G, et al. Noninvasive assessment of pulmonary artery flow and resistance by cardiac magnetic resonance in congenital heart diseases with unrestricted left-to-right shunt. <i>JACC Cardiovasc Imaging</i> . 2009;2(11):1285-1291.	Observational-Dx	26 patients	To determine whether noninvasive assessment of Qp flow by CMR would predict pulmonary vascular resistance in patients with CHD characterized by an unrestricted left-to-right shunt.	ROC analysis was used to determine cutoff values for Qp and Qp/Qs above which the pulmonary vascular resistance could be regarded as clinically acceptable. A Qp of ≥6.05 l/min/m(2) predicted a pulmonary vascular resistance of ≤3.5 WU/m(2) with sensitivity 72%, specificity 100%, and AUC curve 0.90 (P=0.002). A Qp/Qs of ≥2.5/1 predicted a pulmonary vascular resistance of ≤3.5 WU/m(2) with sensitivity 83%, specificity 100%, and AUC 0.94 (P<0.001). Measurement of Qp or left-to-right shunt noninvasively by CMR has potential to predict the pulmonary vascular resistance in patients with an unrestricted left-to-right shunt and could potentially determine operability without having to undertake invasive testing.	3

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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
80. Debl K, Djavidani B, Buchner S, et al. Quantification of left-to-right shunting in adult congenital heart disease: phase-contrast cine MRI compared with invasive oximetry. <i>Br J Radiol.</i> 2009;82(977):386-391.	Observational-Dx	21 patients	To compare phase-contrast cine MRI, a noninvasive imaging technique, with invasive oximetry for the measurement of shunt volumes during cardiac catheterization in adults with left-to-right shunting.	Mean Qp/Qs ratios assessed by phase-contrast cine MRI and oximetry were 2.10+/-0.76 and 1.96+/-0.77, respectively ($P=0.37$). Mean shunt fraction was 46.3+/-19.6% when calculated by phase-contrast cine MRI and 42.3+/-20.1% when obtained by oximetry ($P=0.12$). There was a strong correlation of Qp/Qs ratios and shunt fraction between both methods ($r = 0.61$, $P<0.01$ and $r = 0.84$, $P<0.0001$, respectively). The 2 methods had a good agreement according to Bland and Altman plots with a small but nonsignificant overestimation of Qp/Qs-ratios and shunt fraction by phase-contrast cine MRI. On ROC analysis, the sensitivity and specificity of phase-contrast cine MRI to detect an oximetry-derived Qp/Qs ratio of $\geq 1.5:1$ was 93% and 100% at a phase-contrast cine MRI threshold of a Qp/Qs ratio $\geq 1.75:1$ (AUC = 0.99). Quantification of left-to-right shunting can be performed reliably and accurately by phase-contrast cine MRI and the data obtained by this method correlate closely to those from invasive oximetry.	3
81. Nazarian S, Roguin A, Zviman MM, et al. Clinical utility and safety of a protocol for noncardiac and cardiac magnetic resonance imaging of patients with permanent pacemakers and implantable-cardioverter defibrillators at 1.5 tesla. <i>Circulation.</i> 2006;114(12):1277-1284.	Observational-Dx	55 total patients: 31 pacemaker, 24 implantable defibrillator	To evaluate the diagnostic utility and safety of noncardiac and CMR at 1.5T in patients with implantable cardiac devices using a protocol that incorporates device selection and programming and limits the estimated specific absorption rate of MRI sequences.	Most (66/68 or 97%) of the 68 MRI exams were diagnostic and all were safely performed.	3
82. Bailey WM, Mazur A, McCotter C, et al. Clinical safety of the ProMRI pacemaker system in patients subjected to thoracic spine and cardiac 1.5-T magnetic resonance imaging scanning conditions. <i>Heart Rhythm.</i> 2015:[E-pub ahead of print].	Experimental-Tx	216 patients	To evaluate the clinical safety of the Biotronik ProMRI pacemaker system in patients undergoing thoracic spine and CMR.	In total, 216 patients completed the MRI and 1-month post-MRI follow-up. 1 adverse event possibly related to the implanted system and the MRI procedure occurred, resulting in a serious adverse device effect-free rate of 99.6% (220/221; $P<.0001$). Freedom from atrial and ventricular pacing threshold increase was 100% (194/194, $P<.001$) and 100% (206/206, $P<.001$) respectively. Freedom from P- and R-wave amplitude attenuation was 98.2% (167/170, $P<.001$) and 100% (188/188, $P<.001$) respectively.	2

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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
83. American College of Radiology. <i>Manual on Contrast Media</i> . Available at: http://www.acr.org/Quality-Safety/Resources/Contrast-Manual .	Review/Other-Dx	N/A	Guidance document on contrast media to assist radiologists in recognizing and managing risks associated with the use of contrast media.	N/A	4
84. Farooqi KM, Nielsen JC, Uppu SC, et al. Use of 3-dimensional printing to demonstrate complex intracardiac relationships in double-outlet right ventricle for surgical planning. <i>Circ Cardiovasc Imaging</i> . 2015;8(5):[E-pub ahead of print].	Review/Other-Dx	1 patient	To review the use of 3D printing and modeling in a patient with dextrocardia, complex double-outlet RV and supratricuspid ring to communicate the complex intracardiac anatomy to the surgeon.	No results stated in abstract.	4
85. Kiraly L, Tofeig M, Jha NK, Talo H. Three-dimensional printed prototypes refine the anatomy of post-modified Norwood-1 complex aortic arch obstruction and allow presurgical simulation of the repair. <i>Interact Cardiovasc Thorac Surg</i> . 2015:[E-pub ahead of print].	Review/Other-Dx	1 patient	To present a case of a 5-month old infant with complex obstruction at the neo-aortic to transverse arch and descending aortic junction following the neonatal modified Norwood-1 procedure for hypoplastic left heart syndrome using digital 3D models for insight and simulation.	The solid model provided further insights into details of the anatomy, whereas the surgical approach and steps of the operation were simulated on the hollow model. Intraoperative assessment confirmed the anatomical accuracy of the 3D models. The operation was performed in accordance with preoperative simulation: sliding autologous flaps achieved relief of the obstruction without additional patching. Knowledge gained from the models fundamentally contributed to successful outcome and improved patient safety.	4
86. Partington SL, Valente AM. Cardiac magnetic resonance in adults with congenital heart disease. <i>Methodist Debaquey Cardiovasc J</i> . 2013;9(3):156-162.	Review/Other-Dx	N/A	To review CMR imaging considerations of 3 specific conotruncal congenital heart lesions: tetralogy of Fallot, transposition of the great arteries, and physiologically corrected transposition of the great arteries.	No results stated in abstract.	4
87. Tsai SF, Trivedi M, Daniels CJ. Comparing imaging modalities for screening aortic complications in patients with bicuspid aortic valve. <i>Congenit Heart Dis</i> . 2012;7(4):372-377.	Observational-Dx	106 patients	To compare TTE and CMR imaging for aortic evaluation in patients with bicuspid aortic valve.	There were 106 patients with mean age at CMR 34 +/- 13 years. Mean CMR MaxD was 37 +/- 7 mm. TTE and CMR MaxD mean difference (-1.6 mm) was statistically significant ($P=.002$), particularly when TTE AscAo was not measured (-2.0 mm, $P=.007$). TTE sensitivity was 75% (SV) and 47% (AscAo) for dilation, and 100% (SV) and 83% (AscAo) for aneurysm. Bivariate correlation showed significant positive association between MaxD and diastolic blood pressure and weight ($P<.05$). With multivariate regression, MaxD was significantly smaller in patients with coarctation of the aorta ($P<.001$).	3

* See Last Page for Key

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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
88. Valente AM, Cook S, Festa P, et al. Multimodality imaging guidelines for patients with repaired tetralogy of fallot: a report from the American Society of Echocardiography: developed in collaboration with the Society for Cardiovascular Magnetic Resonance and the Society for Pediatric Radiology. <i>J Am Soc Echocardiogr.</i> 2014;27(2):111-141.	Review/Other-Dx	N/A	To describe the role of each diagnostic modality in the care of patients with repaired tetralogy of fallot and to provide guidelines for a multimodality approach that takes into account patient-related and modality-related considerations.	No results stated in abstract.	4
89. Weber C, Weber M, Ekinci O, et al. Atrial septal defects type II: noninvasive evaluation of patients before implantation of an Amplatzer Septal Occluder and on follow-up by magnetic resonance imaging compared with TEE and invasive measurement. <i>Eur Radiol.</i> 2008;18(11):2406-2413.	Observational-Dx	60 patients	To evaluate morphological and functional MRI of ASDs before and after interventional occlusion by the Amplatzer Septal Occluder in comparison to TEE, invasive balloon measurement and cardiac catheterization.	In MRI, pulmonary-to-systemic flow ratio (Qp/Qs) was calculated and compared with the cardiac catheterization Qp/Qs ratio. Qp/Qs ratio in baseline MRI examination was 1.56 +/- 0.29 (range: 1.05-2.2) and in cardiac catheterization 1.71 +/- 0.30 (range: 1.2-2.4) with a significant correlation (R = 0.65, P<0.01). Defect size on MRI was 15.3 +/- 7.4 mm (range: 3-30 mm), in TEE 14.3 +/- 4.9 mm (range: 4-24 mm), and the balloon stretched diameter in invasive balloon measurement was 23.4 +/- 4.2 mm (range: 14-32 mm). Correlation between defect size in MRI vs TEE was R = 0.67 (P<0.01) and MRI vs invasive balloon measurement was R = 0.77 (P<0.01). RV volumes decreased after intervention.	3
90. Grewal J, Majdalany D, Syed I, Pellikka P, Warnes CA. Three-dimensional echocardiographic assessment of right ventricular volume and function in adult patients with congenital heart disease: comparison with magnetic resonance imaging. <i>J Am Soc Echocardiogr.</i> 2010;23(2):127-133.	Observational-Dx	25 patients	To evaluate the accuracy of 3D US compared with the standard MRI method in determining RV volumes and function in adult patients with CHD and chronic, severe pulmonary regurgitation.	3D US was comparable with MRI in determining RV size and function in most patients with complex CHD. It will be important to study 3D US in a larger population of patients with tetralogy of Fallot, which will be possible only through multi-center collaboration.	2

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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
91. Margossian R, Schwartz ML, Prakash A, et al. Comparison of echocardiographic and cardiac magnetic resonance imaging measurements of functional single ventricular volumes, mass, and ejection fraction (from the Pediatric Heart Network Fontan Cross-Sectional Study). <i>Am J Cardiol.</i> 2009;104(3):419-428.	Observational-Dx	546 total patients: 100 echocardiograms, 50 CMR studies, 124 subjects with paired studies	To compare echocardiographic and CMR measurements of functional single ventricular volumes, mass, and EF.	Volumes by echocardiography averaged 70% of CMR values. Interobserver reproducibility for the EF was similar for the 2 modalities. Although the absolute mean difference between modalities for the EF was small (<2%), 95% limits of agreement were wide. In conclusion, agreement between observers of qualitative functional single ventricular function by echocardiography is modest. Measurements of functional single ventricular volume by 2D echocardiography underestimate CMR measurements, but their reproducibility is high. Echocardiographic and CMR measurements of functional single ventricular EF demonstrate similar interobserver reproducibility, whereas measurements of functional single ventricular mass and LV diastolic volume are more reproducible by CMR.	2
92. Grosse-Wortmann L, Al-Otay A, Yoo SJ. Aortopulmonary collaterals after bidirectional cavopulmonary connection or Fontan completion: quantification with MRI. <i>Circ Cardiovasc Imaging.</i> 2009;2(3):219-225.	Observational-Dx	24 MRI studies	To quantify APC flow after bidirectional cavopulmonary connections and Fontan completions, using phase-contrast MRI, and to identify risk factors for the development of APCs.	The ratio of Qp/Qs was 0.93+/-0.26 in group A and 1.27+/-0.16 in group B. APC flow was 1.42 (0.58 to 3.83) L/min/m(2) and 0.82 (0.50 to 1.81) L/min/m(2) in groups A and B, respectively. The mean inaccuracies corresponded to 7.9+/-14.5% and 7.1+/-13.6% of ascending aortic flow in groups A and B, respectively. Qp/Qs was negatively correlated with a younger age at the time of the bidirectional cavopulmonary connections operation (r=0.62, P=0.01) and positively correlated with the age at the time of the Fontan completion (r=0.81, P=0.01). Patients with a previous right-sided modified Blalock-Taussig shunt had more collateral flow to the right lung than those without.	3

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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
93. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease: Executive Summary: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to develop guidelines for the management of adults with congenital heart disease). <i>Circulation</i> . 2008;118(23):2395-2451.	Review/Other-Dx	N/A	To assist healthcare providers in clinical decision making by describing a range of generally acceptable approaches for diagnosis, management, and prevention of specific diseases or conditions.	No results stated in abstract.	4
94. Cheatham JP, Hellenbrand WE, Zahn EM, et al. Clinical and hemodynamic outcomes up to 7 years after transcatheter pulmonary valve replacement in the US melody valve investigational device exemption trial. <i>Circulation</i> . 2015;131(22):1960-1970.	Experimental-Tx	148 patients	To analyze midterm hemodynamic and clinical outcomes in the IDE trial patients, who were all at least 4 years out from Melody valve implantation.	During a median follow-up of 4.5 years (range, 0.4–7 years), 32 patients underwent RV outflow tract reintervention for obstruction (n=27, with stent fracture in 22), endocarditis (n=3, 2 with stenosis and 1 with pulmonary regurgitation), or RV dysfunction (n=2). 11 patients had the transcatheter pulmonary valve explanted as an initial or second reintervention. 5-year freedom from reintervention and explantation was 76+/-4% and 92+/-3%, respectively. A conduit present and lower discharge RV outflow tract gradient were associated with longer freedom from reintervention. In the 113 patients who were alive and reintervention free, the follow-up gradient (median, 4.5 years after implantation) was unchanged from early post- transcatheter pulmonary valve replacement, and all but 1 patient had mild or less pulmonary regurgitation. Almost all patients were in New York Heart Association class I or II. More severely impaired baseline spirometry was associated with a lower likelihood of improvement in exercise function after transcatheter pulmonary valve replacement.	2

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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
95. Masura J, Gao W, Gavora P, et al. Percutaneous closure of perimembranous ventricular septal defects with the eccentric Amplatzer device: multicenter follow-up study. <i>Pediatr Cardiol.</i> 2005;26(3):216-219.	Experimental-Tx	186 patients	To report our multicenter experience with the Amplatzer eccentric perimembranous ventricular septal defects occluder, focusing on closure rate and ECG changes after implantation.	The device was successfully implanted in all patients. The immediate closure rate was 90% in the first group, increasing to 100% at 1 month and remained at that level during follow-up. The immediate closure rate in the second group was 98% and remained the same during follow-up. The immediate closure rate in the third group was 89% and during 1 year of follow-up remained the same. There was no clinical evidence of hemolysis and no incidence of device embolization or bacterial endocarditis after implantation. Before the procedure, all patients showed normal ECG or LV enlargement. After the procedure (at least 3 months later) ECG showed left anterior hemiblock in 9 patients, complete right bundle branch block in 8 patients, and incomplete right bundle branch block in 7 patients. A complete heart block developed in 2 patients after the procedure (1.07%). The first patient developed left anterior hemiblock immediately after closure and complete heart block within 24 hours, The heart rate was 28 beats per minute. After treatment with steroids and atropine, complete heart block changed to sinus rhythm with left anterior hemiblock within 2 months. 1-year later, the ECG revealed the same findings. The second patient developed complete heart block immediately after the procedure and was on temporary pacing for 1 week. After 1 month, the patient recovered to sinus rhythm and ECG showed left anterior hemiblock.	2

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EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
96. Pass RH, Hijazi Z, Hsu DT, Lewis V, Hellenbrand WE. Multicenter USA Amplatzer patent ductus arteriosus occlusion device trial: initial and one-year results. <i>J Am Coll Cardiol.</i> 2004;44(3):513-519.	Experimental-Tx	439 patients	To review and report initial and 1-year efficacy and safety results of the multicenter USA Amplatzer ductal occluder device trial.	The median patent ductus arteriosus minimal diameter was 2.6 mm (range 0.9 to 11.2 mm); 76 (17%) of 439 were larger than 4.0 mm. Median pulmonary artery mean pressure was 20 mm Hg (range 7 to 80 mm Hg). The Amplatzer ductal occluder was implanted successfully in 435 (99%) of 439 patients, with a median fluoroscopy time of 7.1 min (range 2.9 to 138.4 min). Angiographic demonstration of occlusion was seen in 329 (76%) of 435. This increased to 384 (89%) of 433 on post-catheterization day 1, with occlusion documented in 359 (99.7%) of 360 at 1 year. At the last evaluation in all patients at any time, patent ductus arteriosus closure was documented in 428 (98%) of 435 patients. There have been 2 cases of partial left pulmonary artery occlusion after Amplatzer ductal occluder implantation and no cases of significant aortic obstruction.	2

Evidence Table Key

Study Quality Category Definitions

- *Category 1* The study is well-designed and accounts for common biases.
- *Category 2* The study is moderately well-designed and accounts for most common biases.
- *Category 3* There are important study design limitations.
- *Category 4* The study is not useful as primary evidence. The article may not be a clinical study or the study design is invalid, or conclusions are based on expert consensus. For example:
 - a) the study does not meet the criteria for or is not a hypothesis-based clinical study (e.g., a book chapter or case report or case series description);
 - b) the study may synthesize and draw conclusions about several studies such as a literature review article or book chapter but is not primary evidence;
 - c) the study is an expert opinion or consensus document.
- M = Meta-analysis

Dx = Diagnostic

Tx = Treatment

Abbreviations Key

ACHD = Adults with congenital heart disease

APCs = Aortopulmonary collaterals

ASD = Atrial septal defect

AUC = Area under the curve

CAD = Coronary artery disease

CCTA = Coronary computed tomography angiography

CHD = Congenital heart disease

CI = Confidence interval

CMR = Cardiac magnetic resonance

CT = Computed tomography

CTA = Computed tomography angiography

ECG = Electrocardiogram

EF = Ejection fraction

IV = Intravenous

LV = Left ventricular

MDCT = Multidetector computed tomography

MRA = Magnetic resonance angiography

MRI = Magnetic resonance imaging

OR = Odds ratio

Qp/Qs = pulmonary artery (Qp) and ascending aorta (Qs) ratio

ROC = Receiver-operator characteristic

RV = Right ventricular

SCr = Serum creatinine

TEE = Transesophageal echocardiography

US = Ultrasound