Cardiology in the Young

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Standing on the shoulders of Giants: a citation analysis of the paediatric congenital heart disease literature

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Original Article

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Twitter: Study reveals 100 most cited papers in congenital heart disease, highest cited by Dr Julien Hoffman @UCSFChildrens & Dr Samuel Kaplan @UCLAHealth, most papers in top 100 Dr Jane Newburger @BostonChildrens.

Abstract

Objective: The citation history of a published article reflects its impact on the literature over time. We conducted a comprehensive bibliometric analysis to identify the most cited papers on CHD in children. *Methods:* One-hundred and ninety journals listed in Journal Citation Reports were accessed via Web of Science. Publications with 250 or more citations were identified from Science Citation Index Expanded (1900–2020), and those relating to structural CHD in children were reviewed. Articles were ranked by citation count and the 100 most cited were analysed. *Results:* The number of citations ranged from 2522 to 309 (median 431, IQR 356–518), with 35 published since 2000. All were written in English, most originated from the United States (74%), and were published in cardiovascular journals, with Circulation (28%) the most frequent. There were 86 original research articles, including 50 case series, 14 cohort studies, and 10 clinical trials. The most cited paper was by Hoffman JI and Kaplan S on the incidence of CHD. Thirteen authors had 4 or more publications in the top 100, all of whom had worked in Boston, Philadelphia, San Francisco, or Dallas, and the most prolific author was Newburger JW (9 articles). *Conclusions:* Citation analysis provides a historical perspective on scientific progress by assessing the impact of individual articles. Our study highlights the dominant position of US-based researchers and journals in this field. Most of the highly cited articles remain case series, with few randomised controlled trials in CHD appearing in recent years.

Over the last century, the diagnosis and management of children with CHD has changed rapidly, founded on the work of visionary pioneers, and improved incrementally through advances in knowledge, technique, and technology. Developments in paediatric cardiology, cardiac surgery, imaging, intensive care, genetics, and other areas have radically altered the natural history of CHD, especially for those needing intervention at an early stage in their life trajectory who have the most to gain. Today, we can offer the prospect of "cure" or successful palliation in almost all structural congenital heart conditions.

Progress can be plotted through landmark articles that have made a major impact on the literature,[1,2] providing an insight into the evolution of a specialty.[3] We therefore conducted a comprehensive bibliometric analysis to identify the most cited papers in the field of structural CHD in children.

Methods

Journals listed in the 2019 Journal Citation Reports of the Web of Science (Clarivate Analytics, Philadelphia, PA) under the following subject categories were evaluated for inclusion: Cardiac & Cardiovascular Systems (138 journals); Critical Care Medicine (12); Genetics & Heredity (9); Medicine, General & Internal (14); Multidisciplinary Science (4); Pediatrics (13); Respiratory System (4); Surgery (11); and Transplantation (11). To identify the most cited articles, each journal was searched individually using the Science Citation Index Expanded (1900–2020) accessed via the Web of Science portal on 16 May, 2020. Articles identified by each journal search were ranked according to the number of times cited, from highest to lowest. An additional search was conducted using key words on CHD as the topic or title (see supplement).

Publications with 250 or more citations were identified, and the abstract and/or full text screened independently by both authors to determine whether it was relevant to structural CHD in children; articles in any language were included and any discrepancies resolved by discussion. Articles containing only non-human species, adult patients, or focusing primarily on arrhythmias, vascular disease, normal cardiac function, anesthesia, cardiomyopathies, or primary pulmonary hypertension were excluded. Relevant articles were ranked according to their citation count and the 100 most cited papers were included in the analysis. Data were extracted independently by both authors from the full-text publication, sourced online or via national

libraries, including the title, journal, year, authors, centre and country of origin, language, type of study, level of evidence,[4] field of study, topic, and funding source.

Results

One hundred and ninety journals listed in one or more categories of the Journal Citation Reports were included in the search (see supplement) and 134 articles with 250 or more citations which met the criteria were identified. Articles were ranked according to their citation count and the 100 most cited papers, published in 24 journals, were included in the analysis (Table 1).

The number of citations ranged from 2522 to 309 (median 431, IQR 356–518) and articles were published between 1939 and 2014, with the most cited paper originating from 2002. The most represented decade was the 2000s (28) followed by the 1990s (20), with 35 published since 2000 (Fig 1). All the top 100 papers were published in the English language and the most frequent journals were *Circulation* (28), *Journal of the American College of Cardiology* (10), *New England Journal of Medicine* (8), and *Journal of Pediatrics* and *Journal of Thoracic and Cardiovascular Surgery* (both 7); there were seven papers in non-clinical scientific journals, including four in *Nature*. Most articles originated from the United States (74), followed by Canada (10), United Kingdom (7), Japan (5), France (4), Germany, Italy, and Sweden (2 each), and one each from Austria, the Netherlands, and New Zealand, including seven international collaborations.

There were 542 individuals listed as authors, of whom 81 appeared more than once. Thirteen authors had four or more publications included in the list, as shown in Table 2; all these authors were based at four leading US centres for most or all their publications: Boston Children's Hospital and Harvard University; Children's Hospital of Philadelphia and University of Pennsylvania; University of California San Francisco; and/or University of Texas Southwestern Medical Center. The leading paediatric cardiologists were Dr Jane W Newburger (9) and Dr Gil Wernovsky (8), and the leading surgeons were Dr Richard A Jonas (6), Dr Thomas L Spray, and Dr J William Gaynor (5 each). Fifty-nine papers reported specific funding, with the US National Institutes of Health cited as the sole or a contributing funder in 28 articles.

CHD overall was the most considered disease group (46 articles), followed by the spectrum of single-ventricle heart conditions (11), transposition (8), septal defects (7), and patent arterial duct (6). The most common subject was surgical outcomes (19), followed by pathology (18), surgical technique (13), epidemiology (12), and catheter intervention (9) (Table 3). There were 86 original research articles, including 50 case series, 14 cohort studies, and 10 clinical trials (Table 4); one additional case series was published as a conference abstract. The level of evidence of articles by decade of publication is shown in Figure 2, with level 4 evidence predominant before the 1980s and level 3 evidence most prevalent in more recent decades; all bar one of the most cited level 1 evidence has been published in the last 30 years.

Discussion

The number of times that a published article is cited in the literature reflects its influence and impact over time.[1] Citation counts are being used increasingly as a tool to assess, rank, and compare individuals, institutions, and journals through metrics such as h-index and impact factor.[5] These measures assume that influential researchers and important publications are cited proportionately more often than those of lesser value, providing a measure of impact on the scientific field over time. There is marked variability in the rate of accumulating citations amongst highly cited papers with some peaking early before falling away, whilst others have a long rise to prominence.[2] In this study, we identified the most cited papers on CHD in children, spanning 75 years, from Robert Gross and John Hubbard's landmark 1939 study on surgical ligation of a patent arterial duct in Boston, MA,[6] to Ariane Marelli and colleagues' 2014 paper on the lifetime prevalence of CHD in Quebec, Canada.[7] Whilst many historical classics remain relevant today, one in three of the most highly cited articles was published in the last 20 years.

Types of highly cited study

Much of modern clinical practice in paediatric cardiology and cardiac surgery is based on expert opinion and institutional case series,[8] as reflected by the presence of 61 case reports or series amongst the most cited. Many of the older articles were the classic descriptions of syndromes, such as Williams (727 citations), Patau (563), Cantrell (504), Edwards (500), Alagille (475), and Noonan (408). Similarly, seminal reports of eponymous operations or interventions featured prominently, including Fontan (1631 citations), Blalock and Taussig (837), Mustard (527), Rashkind (523), Senning (490), Norwood (473), and Ross (316). Whilst these procedures remain relevant today and continue to attract attention, it is unlikely that such reports published today would have a similar impact.

Twelve of the top 100 articles were epidemiology studies, including 5 of the top 20 with over 5700 citations between them. Incidence, prevalence, and natural history are often quoted by authors in their introduction as a way of setting the scene, leading to high numbers of citations for those studies that break through and become the standard reference.

Only nine articles reported the outcomes of randomised controlled trials and five of these were on the early and late findings from the Boston Circulatory Arrest trial,[9–13] totalling 2294 citations and involving 8 of the 12 authors appearing in Table 2. The other highly cited randomised controlled trials were the Pediatric Heart Network's multicentre Single-Ventricle Reconstruction trial (463 citations),[14] a national trial of indomethacin in pre-mature infants with a patent arterial duct (427),[15] the multicentre PRIMACORP study of prophylactic milrinone after surgery (404),[16] and the first clinical application of remote ischemic pre-conditioning for myocardial protection (387).[17] The low numbers of randomised controlled trials on the list may reflect the lack of late-phase, practice changing trials in paediatric cardiology and cardiac surgery,[18,19] or that clinical trials address specific research questions, and therefore the findings have a narrower appeal. However, all bar one of these trials were published in the last 30 years yet have garnered sufficient citations to enter the top 100 most cited articles in the field.

Over recent years, clinical guidelines endorsed by national or international bodies have emerged as a key resource for patient management. Four scientific statements from the American Heart Association were included on the list, and it is likely that the number of such articles amongst the most cited will increase in the coming decade.

Origins of highly cited studies

We found that the most cited articles on structural CHD in children were all published in English and dominated by clinical

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Table 1. The top 100 most cited publications in structural CHD in children

Rank	Publication	Citations
1	Hoffman JI, Kaplan S. The incidence of congenital heart disease. *J Am Coll Cardiol* 2002;39(12):1890–900.	2522
2	Fontan F, Baudet E. Surgical repair of tricuspid atresia. *Thorax* 1971;26(3):240–8.	1631
3	Hagen PT, Scholz DG, Edwards WD. Incidence and size of patent foramen ovale during the first 10 decades of life: an autopsy study of 965 normal hearts. *Mayo Clin Proc* 1984;59(1):17–20.	1540
4	Van der Linde D, Konings EEM, Slager MA, Witsenburg M, Helbing WA, Takkenberg JJM, Roos-Hesselink JW. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. *J Am Coll Cardiol* 2011;58(21):2241–7.	990
5	Gatzoulis MA, Balaji S, Webber SA, Siu SC, Hokanson JS, Poile C, Rosenthal M, Nakazawa M, Moller JH, Gillette PC, Webb GD, Redington AN. Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study. *Lancet* 2000;356(9234):975–81.	909
6	Schott JJ, Benson DW, Basson CT, Pease W, Silberbach GM, Moak JP, Maron BJ, Seidman CE, Seidman JG. Congenital heart disease caused by mutations in the transcription factor NKX2-5. *Science* 1998;281(5373):108–11.	874
7	Marelli A, Mackie A, Ionescu-Ittu R, Rahme E, Pilote L. Congenital heart disease in the general population: changing prevalence and age distribution. *Circulation* 2007;115(2):163–72.	869
8	Blalock A, Taussig HB. The surgical treatment of malformations of the heart in which there is pulmonary stenosis or pulmonary atresia. *JAMA* 1945;128(3):189–202.	837
9	Jenkins KJ, Gauvreau K, Newburger JW, Spray TL, Moller JH, Iezzoni LI. Consensus-based method for risk adjustment for surgery for congenital heart disease. *J Thorac Cardiovasc Surg* 2002;123(1):110–8.	799
10	Garg V, Kathiriyra IS, Barnes R, Schluterman MK, King IN, Butler CA, Rothrock CR, Eapen RS, Hirayama-Yamada K, Joo K, Matsuoka R, Cohen JC, Srivastava D. GATA4 mutations cause human congenital heart defects and reveal an interaction with TBX5. *Nature* 2003;424(6947):443–7.	769
11	Mitchell S, Korones S, Berendes H. Congenital heart disease in 56,109 births. Incidence and natural history. *Circulation* 1971;43(3):323–32.	759
12	de Leval MR, Kilner P, Gewillig M, Bull C. Total cavopulmonary connection: a logical alternative to atriopulmonary connection for complex Fontan operations. Experimental studies and early clinical experience. *J Thorac Cardiovasc Surg* 1988;96(5):682–95.	728
13	Williams JC, Barratt-Boyes BG, Lowe JB. Supravalvular aortic stenosis. *Circulation* 1961;24(6):1311–8.	727
14	Basso C, Maron BJ, Corrado D, Thiene G. Clinical profile of congenital coronary artery anomalies with origin from the wrong aortic sinus leading to sudden death in young competitive athletes. *J Am Coll Cardiol* 2000;35(6):1493–501.	654
15	Crafoord C, Nylin G. Congenital coarctation of the aorta and its surgical treatment. *J Thorac Surg* 1945;14:347–361.	643
16	Wernovsky G, Wypij D, Jonas RA, Mayer JE Jr, Hanley FL, Hickey PR, Walsh AZ, Chang AC, Castañeda AR, Newburger JW, Wessel DL. Postoperative course and hemodynamic profile after the arterial switch operation in neonates and infants. A comparison of low-flow cardiopulmonary bypass and circulatory arrest. *Circulation* 1995;92(8):2226–35.	630
17	Murphy JG, Gersh BJ, Mair DD, Fuster V, McGoon MD, Ilstrup DM, McGoon DC, Kirklin JW, Danielson GK. Long-term outcome in patients undergoing surgical repair of tetralogy of Fallot. *N Engl J Med* 1993;329(9):593–9.	616
18	Roberts W. The congenitally bicuspid aortic valve. A study of 85 autopsy cases. *Am J Cardiol* 1970;26(1):72–83.	603
19	Ferencz C, Rubin JD, McCarter RJ, Brenner JI, Neill CA, Perry LW, Hepner SI, Downing JW. Congenital heart disease: prevalence at livebirth. The Baltimore-Washington Infant Study. *Am J Epidemiol* 1985;121(1):31–6.	597
20	Patau K, Smith DW, Therman E, Inhorn SL, Wagner HP. Multiple congenital anomaly caused by an extra autosome. *Lancet* 1960;275(7128):790–3.	563
21	Pagon RA, Graham JM Jr, Zonana J, Yong SL. Coloboma, congenital heart disease, and choanal atresia with multiple anomalies: CHARGE association. *J Pediatr* 1981;99(2):223–7.	536
22	Smith DW, Lemli L, Opitz JM. A newly recognized syndrome of multiple congenital anomalies. *J Pediatr* 1964;64(2):210–7.	532
23	Marino BS, Lipkin PH, Newburger JW, Peacock G, Gerdes M, Gaynor JW, Mussatto KA, Uzark K, Goldberg CS, Johnson WH Jr, Li J, Smith SE, Bellinger DC, Mahle WT. Neurodevelopmental outcomes in children with congenital heart disease: evaluation and management: a scientific statement from the American Heart Association. *Circulation* 2012;126(9):1143–72.	529
24	Mustard WT. Successful two-stage correction of transposition of the great vessels. *Surgery* 1964;55:469–72.	527
25	Rashkind WJ, Miller WW. Creation of an atrial septal defect without thoracotomy. A palliative approach to complete transposition of the great arteries. *JAMA* 1966;196(11):991–2.	523
26	Pierpont ME, Basson CT, Benson DW Jr, Gelb BD, Giglia TM, Goldmuntz E, McGee G, Sable CA, Srivastava D, Webb CL. Genetic basis for congenital heart defects: current knowledge: a scientific statement from the American Heart Association Congenital Cardiac Defects Committee, Council on Cardiovascular Disease in the Young: endorsed by the American Academy of Pediatrics. *Circulation* 2007;115(23):3015–38.	516
27	Korenberg JR, Chen XN, Schipper R, Sun Z, Gonsky R, Gerwehr S, Carpenter N, Daumer C, Dignan P, Disteche C, Graham JM Jr, Hugdins L, McGillivray B, Miyazaki K, Ogasawara N, Park JP, Pagon R, Pueschel S, Sack G, Say B, Schuffenhauer S, Soukup S, Yamakaka T. Down syndrome phenotypes: the consequences of chromosomal imbalance. *Proc Natl Acad Sci USA* 1994;91(11):4997–5001.	512

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Table 1. (*Continued*)

Rank	Publication	Citations
28	Khairy P, Fernandes SM, Mayer JE Jr, Triedman JK, Walsh EP, Lock JE, Landzberg MJ. Long-term survival, modes of death, and predictors of mortality in patients with Fontan surgery. *Circulation* 2008;117(1):85–92.	510
29	Cantrell JR, Haller JA, Ravitch MM. A syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium, and heart. *Surg Gynecol Obstet* 1958;107(5):602–14.	504
30	Edwards JH, Harnden DG, Cameron AH, Crosse VM, Wolff OH. A new trisomic syndrome. *Lancet* 1960;275(7128):787–90.	500
31	Liberthson RR, Sagar K, Berkoben JP, Weintraub RM, Levine FH. Congenital coronary arteriovenous fistula. Report of 13 patients, review of the literature and delineation of management. *Circulation* 1979;59(5):849–54.	497
32	Senning A. Surgical correction of transposition of the great vessels. *Surgery* 1959;45:966–80.	490
32	Taylor AJ, Rogan KM, Virmani R. Sudden cardiac death associated with isolated congenital coronary artery anomalies. *J Am Coll Cardiol* 1992;20(3):640–7.	490
34	Gross RE, Hubbard JP. Surgical ligation of a patent ductus arteriosus: report of first successful case. *JAMA* 1939;112(8):729–31.	489
35	Bellinger DC, Jonas RA, Rappaport LA, Wypij D, Wernovsky G, Kuban KC, Barnes PD, Holmes GL, Hickey PR, Strand RD, Walsh AZ, Helmers SL, Constantinou JE, Carrazana EJ, Mayer JE Jr, Hanley FL, Castaneda AR, Ware JH, Newburger JW. Developmental and neurologic status of children after heart surgery with hypothermic circulatory arrest or lowfFlow cardiopulmonary bypass. *N Engl J Med* 1995;332(9):549–55.	482
36	Morris CA, Demsey SA, Leonard CO, Dilts C, Blackburn BL. Natural history of Williams syndrome: physical characteristics. *J Pediatr* 1988;113(2):318–26.	480
37	Alagille D, Odièvre M, Gautier M, Dommergues JP. Hepatic ductular hypoplasia associated with characteristic facies, vertebral malformations, retarded physical, mental, and sexual development, and cardiac murmur. *J Pediatr* 1975;86(1):63–71.	475
38	Heymann MA, Rudolph AM, Silverman NH. Closure of the ductus arteriosus in premature infants by inhibition of prostaglandin synthesis. *N Engl J Med* 1976;295(10):530–3.	473
38	Norwood WI, Lang P, Hansen DD. Physiologic repair of aortic atresia–hypoplastic left heart syndrome. *N Engl J Med* 1983;308(1):23–6.	473
40	Kan J, White R, Mitchell S, Gardner T. Percutaneous balloon valvuloplasty: a new method for treating congenital pulmonary-valve stenosis. *N Engl J Med* 1982;307(9):540–2.	467
40	Cohen M, Fuster V, Steele PM, Driscoll D, McGoon DC. Coarctation of the aorta. Long-term follow-up and prediction of outcome after surgical correction. *Circulation* 1989;80(4):840–5.	467
42	Ohye RG, Sleeper LA, Mahony L, Newburger JW, Pearson GD, Lu M, Goldberg CS, Tabbutt S, Frommelt PC, Ghanayem NS, Laussen PC, Rhodes JF, Lewis AB, Mital S, Ravishankar C, Williams IA, Dunbar-Masterson C, Atz AM, Colan S, Minich LL, Pizarro C, Kanter KR, Jaggers J, Jacobs JP, Krawczeski CD, Pike N, McCrindle BW, Virzi L, Gaynor JW, Pediatric Heart Network Investigators. Comparison of Shunt Types in the Norwood Procedure for Single-Ventricle Lesions. *N Engl J Med* 2010;362(21):1980–92.	463
43	Bruneau BG. The developmental genetics of congenital heart disease. *Nature* 2008;451(7178):943–8.	451
44	Newburger JW, Jonas RA, Wernovsky G, Wypij D, Hickey PR, Kuban KC, Farrell DM, GL Holmes, Helmers SL, Constantinou J, Carrazana E, Barlow JK, Walsh AZ, Lucius KC, Share JC, Wessel DL, Hanley FL, Mayer JE Jr, Castaneda AR, Ware JH. A comparison of the perioperative neurologic effects of hypothermic circulatory arrest versus low-flow cardiopulmonary bypass in infant heart surgery. *N Engl J Med* 1993;329(15):1057–64.	449
45	Wesselhoeft H, Fawcett JS, Johnson AL. Anomalous origin of the left coronary artery from the pulmonary trunk. Its clinical spectrum, pathology, and pathophysiology based on a review of 140 cases with seven further cases. *Circulation* 1968;38(2):403–25.	443
46	Friedman WF, Hirschklau MJ, Printz MP, Pitlick PT, Kirkpatrick SE. Pharmacologic closure of patent ductus arteriosus in the premature infant. *N Engl J Med* 1976;295(10):526–9.	442
46	Reller MD, Strickland MJ, Riehle-Colarusso T, Mahle WT, Correa A. Prevalence of congenital heart defects in metropolitan Atlanta, 1998–2005. *J Pediatr* 2008;153(6):807–13.	442
48	Du ZD, Hijazi ZM, Kleinman CS, Silverman NH, Larntz K, Amplatzer Investigators. Comparison between transcatheter and surgical closure of secundum atrial septal defect in children and adults: results of a multicenter nonrandomized trial. *J Am Coll Cardiol* 2002;39(11):1836–44.	438
48	Hoffman JIE, Kaplan S, Liberthson RR. Prevalence of congenital heart disease. *Am Heart J* 2004;147(3):425–39.	438
50	Srivastava D, Olson EN. A genetic blueprint for cardiac development. *Nature* 2000;407(6801):221–6.	434
51	Gersony WM, Peckham GJ, Ellison RC, Miettinen OS, Nadas AS. Effects of indomethacin in premature infants with patent ductus arteriosus: results of a national collaborative study. *J Pediatr* 1983;102(6):895–906.	427
51	Jenkins KJ, Correa A, Feinstein JA, Botto L, Britt AE, Daniels SR, Elixson M, Warnes CA, Webb CL, American Heart Association Council on Cardiovascular Disease in the Young. Noninherited risk factors and congenital cardiovascular defects: current knowledge: a scientific statement from the American Heart Association Council on Cardiovascular Disease in the Young: endorsed by the American Academy of Pediatrics. *Circulation* 2007;115(23):2995–3014.	427

(*Continued*)

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Table 1. (*Continued*)

Rank	Publication	Citations
53	Zaidi S, Choi M, Wakimoto H, Ma L, Jiang J, Overton JD, Romano-Adesman A, Bjornson RD, Breitbart RE, Brown KK, Carriero NJ, Cheung YH, Deanfield J, DePalma S, Fakhro KA, Glessner J, Hakonarson H, Italia MJ, Kaltman JR, Kaski J, Kim R, Kline JK, Lee T, Leipzig J, Lopez A, Mane SM, Mitchell LE, Newburger JW, Parfenov M, Pe'er I, Porter G, Roberts AE, Sachidanandam R, Sanders SJ, Seiden HS, State MW, Subramanian S, Tikhonova IR, Wang W, Warburton D, White PS, Williams IA, Zhao H, Seidman JG, Brueckner M, Chung WK, Gelb BD, Goldmuntz E, Seidman CE, Lifton RP. De novo mutations in histone-modifying genes in congenital heart disease. *Nature* 2013;498(7453):220–3.	426
54	Bonnet D, Coltri A, Butera G, Fermont L, Le Bidois J, Kachaner J, Sidi D. Detection of transposition of the great arteries in fetuses reduces neonatal morbidity and mortality. *Circulation* 1999;99(7):916–8.	421
55	Miller SP, McQuillen PS, Hamrick S, Xu D, Glidden DV, Charlton N, Karl T, Azakie A, Ferriero DM, Barkovich AJ, Vigneron DB. Abnormal brain development in newborns with congenital heart disease. *N Engl J Med* 2007;357(19):1928–38.	417
56	Noonan JA. Hypertelorism With Turner phenotype. a new syndrome with associated congenital heart disease. *Am J Dis Child* 1968;116(4):373–80.	408
57	Driscoll DJ, Offord KP, Feldt RH, Schaff HV, Puga FJ, Danielson GK. Five- to fifteen-year follow-up after Fontan operation. *Circulation* 1992;85(2):469–96.	404
57	Hoffman TH, Wernovsky G, Atz AM, Kulik TJ, Nelson DP, Chang AC, Bailey JM, Akbary A, Kocsis JF, Kaczmarek R, Spray TL, Wessel DL. Efficacy and safety of milrinone in preventing low cardiac output syndrome in infants and children after corrective surgery for congenital heart disease. *Circulation* 2003;107(7):996–1002.	404
59	Ohuchi H, Kagisaki K, Miyazaki A, Kitano M, Yazaki S, Sakaguchi H, Ichikawa H, Yamada O, Yagihara T. Impact of the evolution of the Fontan operation on early and late mortality: a single-center experience of 405 patients over 3 decades. *Ann Thorac Surg* 2011;92(4):1457–66.	399
60	Nollert G, Fischlein T, Bouterwek S, Böhmer C, Klinner W, Reichart B. Long-term survival in patients with repair of tetralogy of Fallot: 36-year follow-up of 490 survivors of the first year after surgical repair. *J Am Coll Cardiol* 1997;30(5):1374–83.	396
61	Frieden IJ, Reese V, Cohen D. PHACE syndrome. The association of posterior fossa brain malformations, hemangiomas, arterial anomalies, coarctation of the aorta and cardiac defects, and eye abnormalities. *Arch Dermatol* 1996;132(3):307–11.	387
61	Chessa M, Carminati M, Butera G, Bini RM, Drago M, Rosti L, Giamberti A, Pomè G, Bossone E, Frigiola A. Early and late complications associated with transcatheter occlusion of secundum atrial septal defect. *J Am Coll Cardiol* 2002;39(6):1061–5.	387
63	Cheung MMH, Kharbanda RK, Konstantinov IE, Shimizu M, Frndova H, Li J, Holtby HM, Cox PN, Smallhorn JF, Van Arsdell GS, Redington AN. Randomized controlled trial of the effects of remote ischemic preconditioning on children undergoing cardiac surgery: first clinical application in humans. *J Am Coll Cardiol* 2006;47(11):2277–82.	387
63	Khairy P, Ionescu-Ittu R, Mackie AS, Abrahamowicz M, Pilote L, Marelli AJ. Changing mortality in congenital heart disease. *J Am Coll Cardiol* 2010;56(14):1149–57.	387
65	Bellinger DC, Wypij D, duPlessis AJ, Rappaport LA, Jonas RA, Wernovsky G, Newburger JW. Neurodevelopmental status at eight years in children with dextro-transposition of the great arteries: the Boston Circulatory Arrest Trial. *J Thorac Cardiovasc Surg* 2003;126(5):1385–96.	384
66	Krichenko A, Benson LN, Burrows P, Möes CA, McLaughlin P, Freedom RM. Angiographic classification of the isolated, persistently patent ductus arteriosus and implications for percutaneous catheter occlusion. *Am J Cardiol* 1989;63(12):877–80.	380
67	Collett RW, Edwards JE. Persistent truncus arteriosus: a classification according to anatomic types. *Surg Clin North Am* 1949;29(4):1245–70.	378
68	Simon P, Kasimir MT, Seebacher G, Weigel G, Ullrich R, Salzer-Muhar U, Rieder E, Wolner E. Early failure of the tissue engineered porcine heart valve SYNERGRAFT in pediatric patients. *Eur J Cardiothorac Surg* 2003;23(6):1002–6.	375
69	Tworetzky W, McElhinney DB, Reddy VM, Brook MM, Hanley FL, Silverman NH. Improved surgical outcome after fetal diagnosis of hypoplastic left heart syndrome. *Circulation* 2001;103(9):1269–73.	374
70	Hoffman JI. Incidence of congenital heart disease: I. Postnatal incidence. *Pediatr Cardiol* 1995;16(3):103–13.	372
71	Reifenstein GH, Levine SA, Gross RE. Coarctation of the aorta, a review of 104 autopsied cases of the adult type, 2 years of age or older. *Am Heart J* 1947;33(2):146–68.	366
72	Fontan F, Kirklin JW, Fernandez G, Costa F, Naftel DC, Tritto F, Blackstone EH. Outcome after a "perfect" Fontan operation. *Circulation* 1990;81(5):1520–36.	358
72	Olson EN, Srivastava D. Molecular pathways controlling heart development. *Science* 1996;272(5262):671–6.	358
74	Van Praagh R, Van Praagh S. The anatomy of common aorticopulmonary trunk (truncus arteriosus communis) and its embryologic implications. A study of 57 necropsy cases. *Am J Cardiol* 1965;16(3):406–25.	357
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76	Hoffman JI, Christianson R. Congenital heart disease in a cohort of 19,502 births with long-term follow-up. *Am J Cardiol* 1978;42(4):641–7.	354

(*Continued*)

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Table 1. (*Continued*)

Rank	Publication	Citations
77	Potts WJ, Smith S, Gibson S. Anastomosis of the aorta to a pulmonary artery; certain types in congenital heart disease. *JAMA* 1946;132(11):627–31.	351
77	Lewis FJ, Taufic M. Closure of atrial septal defects with the aid of hypothermia; experimental accomplishments and the report of one successful case. *Surgery* 1953;33(1):52–59.	351
79	Bellinger DC, Wypij D, Kuban KC, Rappaport LA, Hickey PR, Wernovsky G, Jonas RA, Newburger JW. Developmental and neurological status of children at 4 years of age after heart surgery with hypothermic circulatory arrest or low-flow cardiopulmonary bypass. *Circulation* 1999;100(5):526–32.	349
80	Marelli AJ, Ionescu-Ittu R, Mackie AS, Guo L, Dendukuri N, Kaouache M. Lifetime prevalence of congenital heart disease in the general population from 2000 to 2010. *Circulation* 2014;130(9):749–56.	348
81	Nakata S, Imai Y, Takanashi Y, Kurosawa H, Tezuka K, Nakazawa M, Ando M, Takao A. A new method for the quantitative standardization of cross-sectional areas of the pulmonary arteries in congenital heart diseases with decreased pulmonary blood flow. *J Thorac Cardiovasc Surg* 1984;88(4):610–9.	347
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83	O'Laughlin MP, Perry SB, Lock JE, Mullins CE. Use of endovascular stents in congenital heart disease. *Circulation* 1991;83(6):1923–39.	331
84	Tweddell JS, Hoffman GM, Mussatto KA, Fedderly RT, Berger S, Jaquiss RDB, Ghanayem NS, Frisbee SJ, Litwin SB. Improved survival of patients undergoing palliation of hypoplastic left heart syndrome: lessons learned from 115 consecutive patients. *Circulation* 2002;106 (12 Suppl 1):I82–9.	329
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89	Donofrio MT, Moon-Grady AJ, Hornberger LK, Copel JA, Sklansky MS, Abuhamad A, Cuneo BF, Huhta JC, Jonas RA, Krishnan A, Lacey S, Lee W, Michelfelder EC Sr, Rempel GR, Silverman NH, Spray TL, Strasburger JF, Tworetzky W, Rychik J; on behalf of the American Heart Association Adults with Congenital Heart Disease Joint Committee of the Council on Cardiovascular Disease in the Young and Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and Council on Cardiovascular and Stroke Nursing. Diagnosis and treatment of fetal cardiac disease: a scientific statement from the American Heart Association. *Circulation* 2014;129(21):2183–242.	322
90	Rashkind WJ, Mullins CE, Hellenbrand WE, Tait MA. Nonsurgical closure of patent ductus arteriosus: clinical application of the Rashkind PDA Occluder System. *Circulation* 1987;75(3):583–92.	319
90	Amin Z, Hijazi ZM, Bass JL, Cheatham JP, Hellenbrand WE, Kleinman CS. Erosion of Amplatzer septal occluder device after closure of secundum atrial septal defects: review of registry of complications and recommendations to minimize future risk. *Catheter Cardiovasc Interv* 2004;63(4):496–502.	319
92	Ross DN, Somerville J. Correction of pulmonary atresia with a homograft aortic valve. *Lancet* 1966;288(7479):1446–7.	316
93	Conley ME, Beckwith JB, Mancer JF, Tenckhoff L. The spectrum of the DiGeorge syndrome. *J Pediatr* 1979;94(6):883–90.	315
93	Mahle WT, Tavani F, Zimmerman RA, Nicolson SC, Galli KK, Gaynor JW, Clancy RR, Montenegro LM, Spray TL, Chiavacci RM, Wernovsky G, Kurth CD. An MRI study of neurological injury before and after congenital heart surgery. *Circulation* 2002;106(12 Suppl 1):I109–14.	315
93	Licht DJ, Shera DM, Clancy RR, Wernovsky G, Montenegro LM, Nicolson SC, Zimmerman RA, Spray TL, Gaynor JW, Vossough A. Brain maturation is delayed in infants with complex congenital heart defects. *J Thorac Cardiovasc Surg* 2009;137(3):529–36.	315
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96	Lababidi Z, Wu JR, Walls JT. Percutaneous balloon aortic valvuloplasty: results in 23 patients. *Am J Cardiol* 1984;53(1):194–7.	314
98	Celoria GC, Patton RB. Congenital absence of the aortic arch. *Am Heart J* 959;58(3):407–13.	313
99	Niwa K, Perloff JK, Bhuta SM, Laks H, Drinkwater DC, Child JS, Miner PD. Structural abnormalities of great arterial walls in congenital heart disease: light and electron microscopic analyses. *Circulation* 2001;103(3):393–400.	310
100	Rosenzweig EB, Kerstein D, Barst RJ. Long-term prostacyclin for pulmonary hypertension with associated congenital heart defects. *Circulation* 1999;99(14):1858–65.	309

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Table 2. Authors with 4 or more articles in the 100 most cited

Author	Publications (n)
Jane W Newburger, *Boston Children's Hospital and Harvard Medical School*	9
Gil Wernovsky, *Boston Children's Hospital and Harvard Medical School, and Children's Hospital of Philadelphia and University of Pennsylvania*	8
Richard A Jonas, *Boston Children's Hospital and Harvard Medical School, and Children's National Medical Center*	6
J William Gaynor, *Children's Hospital of Philadelphia and University of Pennsylvania*	5
Thomas L Spray, *Children's Hospital of Philadelphia and University of Pennsylvania*	5
David Wypij, *Boston Children's Hospital and Harvard School of Public Health*	5
David C Bellinger, *Boston Children's Hospital and Harvard Medical School*	4
Frank L Hanley, *Boston Children's Hospital and Harvard Medical School, and University of California San Francisco*	4
Paul R Hickey, *Boston Children's Hospital and Harvard Medical School*	4
Julien I E Hoffman, *University of California San Francisco*	4
John E Mayer Jr, *Boston Children's Hospital and Harvard Medical School*	4
Norman H Silverman, *University of California San Francisco, and Stanford University*	4
Deepak Srivastava, *University of Texas Southwestern Medical Center, and University of California San Francisco*	4

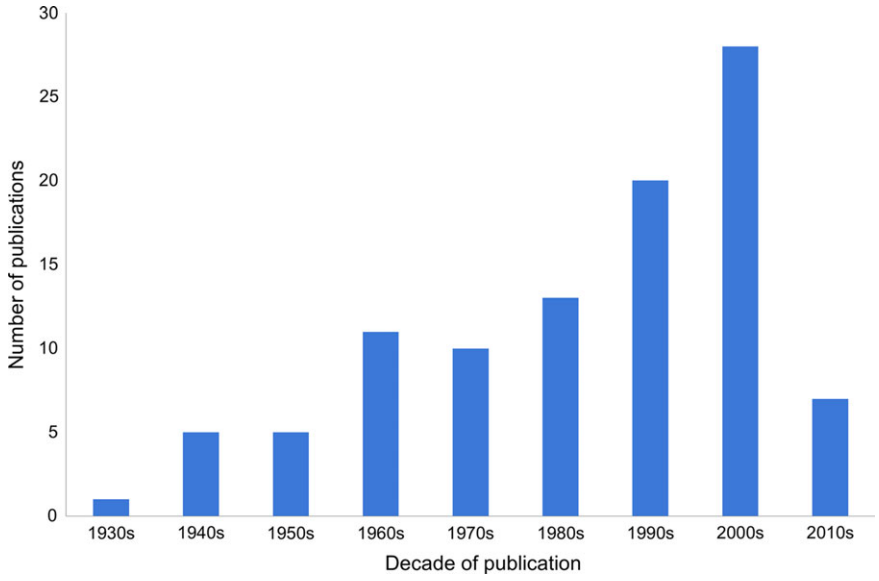

Figure 1. Number of articles in the top 100 most cited, by decade of publication.

research groups in the United States, as found in other areas of cardiovascular disease[20,21]; only 16 involved authors from non-English speaking countries and 4 originated from outside Europe or North America. Most articles were publishing in US-based cardiovascular journals, whilst almost a quarter appeared in high-impact general medical (17) or non-clinical scientific (7) journals. It is not surprising that of those rapidly rising studies published in the last 15 years, 14 of 16 (88%) were published in high-impact specialty or general medical journals.

The observation that all authors featuring four or more times had worked at one of four US institutions for most or all their impactful publications reinforces the benefits of "team science." These groups established and sustained successful and collaborative research programmes, where success breeds success, although multiple highly cited papers originating from one clinical trial contributed much to this achievement. However, it also raises the possibility of a publication bias in which work from leading US institutions is favoured by leading US journals.

The most highly cited papers

At the time of our search, three papers had generated over 1000 citations. Julien Hoffman and Samuel Kaplan's article on the incidence of CHD was by far the most cited with 2,522 citations.[22] Published in 2002, it is likely to retain this position for many years, although the systematic review and meta-analysis on worldwide prevalence by Van der Linde and colleagues from 2011 is rising fast, already in fourth place with 990 citations.[23] The second most cited paper was a surgical classic from 1971, Fontan and Baudet's original description of ventricularisation of the right atrium to direct inferior vena caval blood to the left lung, receiving 1631 citations[24]; the evolution and outcomes of the Fontan

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Table 3. Subjects of studies in the 100 most cited

Subject	Publications (n)
Surgical outcomes	19
Pathology	18
Surgical technique	13
Epidemiology	12
Catheter intervention	9
Genetics	8
Morphology	8
Medical therapy	6
Fetal	4
Surgical risk modelling	2
Imaging	1

Table 4. Types of studies in the 100 most cited

Type of study	Publications (n)
Case series	51
Cohort studies	14
Clinical trials	10
Randomised controlled trial	9
Non-randomised trial	1
Single case reports	10
Reviews	7
Narrative reviews	5
Systematic reviews	2
Scientific statements	4
Expert consensus/opinions	2
Case control study	1
Data modelling	1

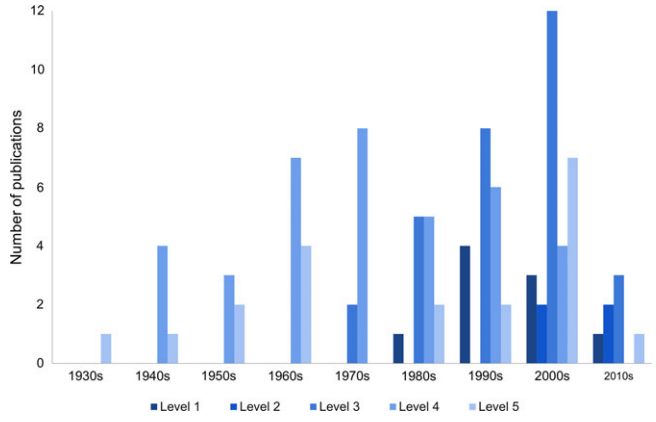

Figure 2. Level of evidence of articles in the top 100 most cited, by decade of publication.

procedure have remained the focus of intense research over the last five decades. In third place was Philip Hagen and colleagues' 1984 autopsy study on the incidence and size of patent foramen ovale by age in children and adults and has grossed over 1500 citations.[25]

Strengths and limitations

This study provides a unique insight into the most influential peer-reviewed publications that have shaped the development of our specialty over the last 100 years, providing a reference list that will be of interest to students, clinicians, and researchers. Our comprehensive search of a wide range of journals listed on the expanded *Science Citation Index* is likely to have missed few if any of the most cited papers. However, our findings are limited by our use of only one bibliometric tool to determine citation counts; other databases, such as *Scopus* or *Google Scholar*, use different data sources and algorithms which may impact on the number of citations detected and therefore which papers appear in the top 100[26] although as both index fewer articles published prior to the 1990s, they are less suited to historical analyses. Similarly, metrics related to other sources of information, such as number of times accessed online and twitter engagements, were not assessed but are much more relevant to recently published works. Citation analysis demonstrates the overall impact of a paper since publication but is not adjusted to account for its current influence or its quality; infamous papers may receive as much attention as celebrated papers. Like any bibliometric analysis, our findings are a snapshot in time, a now historical view on the most cited articles in the field. Since our search, recently published articles, such as Zaidi et al. in *Nature* 2013 (426 citations)[27] and Marelli et al in *Circulation* 2014 (348),[7] will have continued their rapid rise, whilst others, such as the scientific statement by Pierpont et al. in *Circulation* 2007 (516),[28] have been superseded by updated reviews[29] and are therefore likely to plateau. As a time-dependent process, other groundbreaking but recently published articles may have had too little time to accrue sufficient citations to make our list.

Conclusions

This study identified the most influential papers on CHD in children, providing a historical perspective on scientific progress over the last century. Our findings demonstrate that highly cited papers are exclusively published in English, and that researchers and journals based in the United States hold a dominant position in this field. Most of the highly cited articles remain case series, with few randomised controlled trials in CHD appearing in recent years.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951121001256

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Conflicts of interest. None.

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