

Original Article

Citation classics in chronic granulomatous disease: a bibliometric analysis

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Abstract: The number of times a scientific article has been cited reflects its impact on a specific field. Highly cited articles are known as citation classics. Here, we aim to characterize the most frequently cited articles in chronic granulomatous disease (CGD). We searched the ISI Web of Science in Aug 2016 for articles that were cited 100 times or more and evaluated them for several characteristics. The most frequently cited article received 955 citations. The citations mean was 221.6 citations (SD = 162.7). The most recent article was published in 2011. The articles were published in 36 journals, led by The New England Journal of Medicine. Overall authors came from 17 countries, with the United States of America (USA) contributing to 91 (80%) articles. The National Institute of Health was the most common institution of origin for the corresponding author. The most common type of articles was basic science with 77 (67%) articles. These results provide some insights into the most cited research articles in CGD since its first description 60 years ago.

Keywords: Chronic granulomatous disease, immunodeficiency, phagocytosis, aspergillosis, citation classics, top cited, ISI web of science, bibliometrics, top 100, landmark article

Introduction

Chronic granulomatous disease (CGD) is an extremely rare congenital immune deficiency syndrome with a prevalence of approximately 1:250,000 individuals. It is characterized by recurrent severe infections due to the inability of neutrophils and macrophages to mount a respiratory burst and kill invading bacteria and fungi. It is caused by nicotinamide adenine dinucleotide phosphate (NADPH) oxidase deficiency [1].

In 1954, Janeway et al. [2] first recognized CGD in a group of five affected boys. However, since that time, much has been learned about CGD and several milestones in its understanding were achieved [3]. The disease has been in the forefront of what we have learned about anti-bacterial prophylaxis, cytokine immunotherapy, bone marrow transplantation, and gene therapy [4].

Methods to identify milestones in a specific research field include peer review and citation

analysis [5]. The number of times an article is cited in scientific journals measures its impact on a specific biomedical field [6]. In 1987, Garfield listed the “top 100” best cited articles ever published in JAMA and named them citation classics [5]. Since then, there have been several reports identifying citation classics in specific medical fields [6-41] and in specific journals [42-44]. However, while citation analysis has been criticized, it may allow for the identification of advances in a particular specialty and may provide a historical perspective on its scientific progress [7, 45]. Here, we identify and analyze, the citation classics in CGD. To our knowledge, this is the first of such a study in CGD and in any primary immunodeficiency disease.

Materials and methods

We performed a search on the ISI Web of Science for article titles with the keyword “chronic granulomatous disease”, “fatal granulomatous disease of childhood”, “a syndrome of recurrent

Citation classics in CGD

Table 1. Citation classics articles in CGD ranked according to the total number of citations

Rank according to number of citation	Number of citation	Citation rate per year	Article	Year	Journal title	1 st author
1	955	20.32	Quantitative nitroblue tetrazolium test in chronic granulomatous disease.	1968	N Engl J Med	Baehner RL
2	930	19.38	In vitro bactericidal capacity of human polymorphonuclear leukocytes: diminished activity in chronic granulomatous disease of childhood.	1967	J Clin Invest	Quie PG
3	762	50.8	Chronic granulomatous disease. Report on a national registry of 368 patients.	2000	Medicine (Baltimore)	Winkelstein JA
4	733	18.34	The role of superoxide anion generation in phagocytic bactericidal activity. Studies with normal and chronic granulomatous disease leukocytes	1975	J Clin Invest	Johnston RB Jr
5	687	76.33	Correction of X-linked chronic granulomatous disease by gene therapy, augmented by insertional activation of MDS1-EVI1, PRDM16 or SETBP1.	2006	Nat Med	Ott MG
6	664	22.9	Cloning the gene for an inherited human disorder—chronic granulomatous disease—on the basis of its chromosomal location.	1986	Nature	Royer-Pokora B
7	583	29.15	Mouse model of X-linked chronic granulomatous disease, an inherited defect in phagocyte superoxide production.	1995	Nat Genet	Pollock JD
8	522	34.8	Genetic, biochemical, and clinical features of chronic granulomatous disease.	2000	Medicine (Baltimore)	Segal BH
9	421	8.77	Leukocyte oxidase: defective activity in chronic granulomatous disease.	1967	Science	Baehner RL
10	399	13.3	Minor Xp21 chromosome deletion in a male associated with expression of Duchenne muscular dystrophy, chronic granulomatous disease, retinitis pigmentosa, and McLeod syndrome.	1985	Am J Hum Genet	Francke U
11	367	7.49	Fatal granulomatous disease of childhood. An inborn abnormality of phagocytic function.	1966	Lancet	Holmes B
12	356	17.8	The p47 phox mouse knock-out model of chronic granulomatous disease.	1995	J Exp Med	Jackson SH
13	340	8.29	Defective superoxide production by granulocytes from patients with chronic granulomatous disease.	1974	N Engl J Med	Curnutte JT
14	339	67.8	Genomic instability and myelodysplasia with monosomy 7 consequent to EVI1 activation after gene therapy for chronic granulomatous disease.	2010	Nat Med	Stein S
15	336	12.44	Two cytosolic neutrophil oxidase components absent in autosomal chronic granulomatous disease.	1988	Science	Volpp BD
16	335	47.86	Defective tryptophan catabolism underlies inflammation in mouse chronic granulomatous disease.	2008	Nature	Romani L
17	323	12.42	Recombinant 47-kilodalton cytosol factor restores NADPH oxidase in chronic granulomatous disease.	1989	Science	Lomax KJ
18a	310	16.32	Mutations in the X-linked and autosomal recessive forms of chronic granulomatous disease.	1996	Blood	Roos D
18b	310	11.07	The glycoprotein encoded by the X-linked chronic granulomatous disease locus is a component of the neutrophil cytochrome b complex.	1987	Nature	Dinauer MC
19	309	9.09	Fungal infection in chronic granulomatous disease. The importance of the phagocyte in defense against fungi.	1981	Am J Med	Cohen MS
20	298	12.42	A controlled trial of interferon gamma to prevent infection in chronic granulomatous disease. The International Chronic Granulomatous Disease Cooperative Study Group.	1991	N Engl J Med	The international CGD cooperative study group
21	297	12.38	Molecular basis of chronic granulomatous disease.	1991	Blood	Smith RM
22	284	10.52	Two forms of autosomal chronic granulomatous disease lack distinct neutrophil cytosol factors.	1988	Science	Nunoi H
23	282	7.23	The biochemical basis of nitroblue tetrazolium reduction in normal human and chronic granulomatous disease polymorphonuclear leukocytes	1976	Blood	Baehner RL
24	273	10.5	The electron transport chain of the microbicidal oxidase of phagocytic cells and its involvement in the molecular pathology of chronic granulomatous disease.	1989	J Clin Invest	Segal AW

Citation classics in CGD

25	266	9.5	Absence of both cytochrome b-245 subunits from neutrophils in X-linked chronic granulomatous disease.	1987	Nature	Segal AW
26	263	8.22	Absence of cytochrome b-245 in chronic granulomatous disease. A multicenter European evaluation of its incidence and relevance.	1983	N Engl J Med	Segal AW
27	260	10.4	Human neutrophil cytochrome b light chain (p22-phox). Gene structure, chromosomal location, and mutations in cytochrome-negative autosomal recessive chronic granulomatous disease.	1990	J Clin Invest	Dinauer MC
28	256	21.33	Chronic granulomatous disease.	2003	Curr Opin Immunol	Heyworth PG
29	252	8.4	Stimulated neutrophils from patients with autosomal recessive chronic granulomatous disease fail to phosphorylate a Mr-44,000 protein.	1985	Nature	Segal AW
30	243	8.68	The X-linked chronic granulomatous disease gene codes for the beta-chain of cytochrome b-245.	1987	Nature	Teahan C
31	241	7.53	Chronic granulomatous disease: a syndrome of phagocyte oxidase deficiencies.	1983	Medicine (Baltimore)	Tauber AI
32	231	12.83	Absence of respiratory burst in X-linked chronic granulomatous disease mice leads to abnormalities in both host defense and inflammatory response to <i>Aspergillus fumigatus</i> .	1997	J Exp Med	Morgenstern DE
33	227	4.05	A fatal granulomatous disease of childhood; the clinical, pathological, and laboratory features of a new syndrome.	1959	AMA J Dis Child	Bridges RA
34	226	8.37	Partial correction of the phagocyte defect in patients with X-linked chronic granulomatous disease by subcutaneous interferon gamma.	1988	N Engl J Med	Ezekowitz RAB
35	220	3.79	A syndrome of recurrent infection and infiltration of viscera by pigmented lipid histiocytes.	1957	Pediatrics	Landing BH
36	219	12.17	Prolonged production of NADPH oxidase-corrected granulocytes after gene therapy of chronic granulomatous disease.	1997	Proc Natl Acad Sci U S A	Malech HL
37	208	8.0	Incidence, severity, and prevention of infections in chronic granulomatous disease.	1989	J Pediatr	Mouy R
38a	207	5.91	Use of lipophilic probes of membrane potential to assess human neutrophil activation. Abnormality in chronic granulomatous disease.	1980	J Clin Invest	Seligmann BE
38b	207	5.45	Chronic granulomatous disease.	1977	Pediatr Clin North Am	Johnston RB
39	206	6.44	Recent advances in chronic granulomatous disease.	1983	Ann Intern Med	Gallin JI
40a	205	9.76	Chronic granulomatous disease.	1994	Biochim Biophys Acta	Thrasher AJ
40b	205	7.88	Genetic variants of chronic granulomatous disease: prevalence of deficiencies of two cytosolic components of the NADPH oxidase system.	1989	N Engl J Med	Clark RA
41	202	5.05	NADPH oxidase deficiency in X-linked chronic granulomatous disease.	1975	J Clin Invest	Hohn DC
42	199	5.69	Correlation between membrane potential changes and superoxide production in human granulocytes stimulated by phorbol myristate acetate. Evidence for defective activation in chronic granulomatous disease.	1980	J Biol Chem	Whitin JC
43	192	4.17	Iodination defect in the leukocytes of a patient with chronic granulomatous disease of childhood.	1969	N Engl J Med	Klebanoff SJ
44	186	4.23	Chronic granulomatous disease: correlation between pathogenesis and clinical findings.	1971	Pediatrics	Johnston RB Jr
45a	183	10.76	Host defense molecule polymorphisms influence the risk for immune-mediated complications in chronic granulomatous disease.	1998	J Clin Invest	Foster CB
45b	183	6.54	Chronic granulomatous disease.	1987	Adv Hum Genet	Curnutte JT
46a	181	30.17	Chronic granulomatous disease: the European experience.	2009	PLoS One	van den Berg JM
46b	181	6.24	A possible role for protein phosphorylation in the activation of the respiratory burst in human neutrophils. Evidence from studies with cells from patients with chronic granulomatous disease.	1986	J Biol Chem	Hayakawa T
47	179	3.81	Deficiency of reduced nicotinamide-adenine dinucleotide oxidase in chronic granulomatous disease.	1968	Science	Baehner RL

Citation classics in CGD

48	174	3.78	Failure of nitro blue tetrazolium reduction in the phagocytic vacuoles of leukocytes in chronic granulomatous disease.	1969	J Clin Invest	Nathan DG
49	172	6.37	Relationship of protein phosphorylation to the activation of the respiratory burst in human neutrophils. Defects in the phosphorylation of a group of closely related 48-kDa proteins in two forms of chronic granulomatous disease.	1988	J Biol Chem	Okamura N
50	169	12.07	Treatment of chronic granulomatous disease with nonmyeloablative conditioning and a T-cell-depleted hematopoietic allograft.	2001	N Engl J Med	Horwitz ME
51	167	13.92	Itraconazole to prevent fungal infections in chronic granulomatous disease.	2003	N Engl J Med	Gallin JI
52	166	27.67	A new genetic subgroup of chronic granulomatous disease with autosomal recessive mutations in p40 (phox) and selective defects in neutrophil NADPH oxidase activity.	2009	Blood	Matute JD
53a	165	7.5	Gene targeting of X chromosome-linked chronic granulomatous disease locus in a human myeloid leukemia cell line and rescue by expression of recombinant gp91 phox.	1993	Proc Natl Acad Sci U S A	Zhen L
53b	165	3.67	A simple rapid micromethod for detecting chronic granulomatous disease of childhood.	1970	J Lab Clin Med	Gifford RH
53c	165	3.59	Degranulation of leukocytes in chronic granulomatous disease.	1969	J Clin Invest	Baehner RL
54	164	32.8	Chronic granulomatous disease.	2010	Clin Rev Allergy Immuno	Holland SM
55	160	3.56	Chronic granulomatous disease in females.	1970	N Engl J Med	Holmes B
56a	155	31.0	Residual NADPH oxidase and survival in chronic granulomatous disease.	2010	N Engl J Med	Kuhns DB
56b	155	14.09	Gastrointestinal involvement in chronic granulomatous disease.	2004	Pediatrics	Marciano BE
57	154	4.16	Absence of a newly described cytochrome b from neutrophils of patients with chronic granulomatous disease.	1978	Lancet	Segal AW
58	153	3.33	Leukocyte function in chronic granulomatous disease of childhood. Studies on a seventeen year old boy.	1969	Am J Med	Mandell GL
59	150	3.57	Gastrointestinal manifestations of chronic granulomatous disease.	1973	N Engl J Med	Ament ME
60	149	5.52	Recombinant human interferon-gamma reconstitutes defective phagocyte function in patients with chronic granulomatous disease of childhood.	1988	Proc Natl Acad Sci U S A	Sechler JM
61	147	4.08	Prenatal diagnosis of chronic granulomatous disease.	1979	N Engl J Med	Newburger PE
62	145	2.9	Thirteen boys with progressive septic granulomatosis.	1965	Pediatrics	Carson MJ
63a	144	3.43	The NBT slide test: a simple screening method for detecting chronic granulomatous disease and female carriers.	1973	J Pediatr	Ochs HD
63b	144	3.06	Defective polymorphonuclear-leukocyte function and chronic granulomatous disease in two female children.	1968	N Engl J Med	Quie PG
64a	143	8.41	Aspergillus nidulans infection in chronic granulomatous disease.	1998	Medicine (Baltimore)	Segal BH
64b	143	3.49	Effects of phorbol myristate acetate on the metabolism and ultrastructure of neutrophils in chronic granulomatous disease.	1974	J Clin Invest	Repine JE
65	141	3.36	Neutrophil dysfunction, chronic granulomatous disease, and non-spherocytic haemolytic anaemia caused by complete deficiency of glucose-6-phosphate dehydrogenase.	1973	Lancet	Gray GR
66	138	2.94	Studies of polymorphonuclear leukocytes from patients with chronic granulomatous disease of childhood: bactericidal capacity for streptococci.	1968	Pediatrics	Kaplan EL
67	137	4.89	Recombinant interferon gamma augments phagocyte superoxide production and X-linked chronic granulomatous disease gene expression in X-linked variant chronic granulomatous disease.	1987	J Clin Invest	Ezekowitz RA
68	136	10.46	Treatment of chronic granulomatous disease with myeloablative conditioning and an unmodified hemopoietic allograft: a survey of the European experience, 1985-2000.	2002	Blood	Segar RA
69	135	3.14	Phagocytosis in chronic granulomatous disease and the Chediak-Higashi syndrome.	1972	N Engl J Med	Stossel TP

Citation classics in CGD

70	132	5.28	Trimethoprim-sulfamethoxazole prophylaxis in the management of chronic granulomatous disease.	1990	J Infect Dis	Margolis DM
71a	131	2.85	Leukocyte bactericidal activity in chronic granulomatous disease: correlation of bacterial hydrogen peroxide production and susceptibility to intracellular killing.	1969	J Bacteriol	Mandell GL
71b	131	2.79	The pattern of genetic transmission of the leukocyte defect in fatal granulomatous disease of childhood.	1968	J Clin Invest	Windhorst DB
72a	129	32.25	Oxidase-deficient neutrophils from X-linked chronic granulomatous disease iPS cells: functional correction by zinc finger nuclease-mediated safe harbor targeting.	2011	Blood	Zou J
72b	129	4.96	Absence of both the 91 kD and 22 kD subunits of human neutrophil cytochrome b in two genetic forms of chronic granulomatous disease.	1989	Blood	Parkos CA
73	127	18.14	Clinical features, long term follow-up and outcome of a large cohort of patients with chronic granulomatous disease: an Italian multicenter study.	2008	Clin Immunol	Martire B
74	126	2.93	Comparative study of the metabolic and bactericidal characteristics of severely glucose-6-phosphate dehydrogenase-deficient polymorphonuclear leukocytes and leukocytes from children with chronic granulomatous disease.	1972	J Reticuloendothel Soc	Baehner RL
75a	125	4.63	Purified protein kinase C phosphorylates a 47-kDa protein in control neutrophil cytoplasts but not in neutrophil cytoplasts from patients with the autosomal form of chronic granulomatous disease.	1988	J Biol Chem	Kramer IM
75b	125	3.13	Defect in pyridine nucleotide dependent superoxide production by a particulate fraction from the granulocytes of patients with chronic granulomatous disease.	1975	N Engl J Med	Curnutte JT
76	124	4.0	B cell lines as models for inherited phagocytic diseases: abnormal superoxide generation in chronic granulomatous disease and giant granules in Chediak-Higashi syndrome.	1984	J Immunol	Volkman DJ
77	123	24.6	Reactive oxygen species-independent activation of the IL-1 beta inflammasome in cells from patients with chronic granulomatous disease.	2010	Proc Natl Acad Sci U S A	Van der Veerdonk FL
78	122	4.21	Neutrophil-mediated solubilization of the subendothelial matrix: oxidative and nonoxidative mechanisms of proteolysis used by normal and chronic granulomatous disease phagocytes.	1986	J Immunol	Weiss SJ
79	120	5.71	Infection with Pseudomonas cepacia in chronic granulomatous disease: role of nonoxidative killing by neutrophils in host defense.	1994	J Infect Dis	Speert DP
80	119	4.58	A missense mutation in the neutrophil cytochrome b heavy chain in cytochrome-positive X-linked chronic granulomatous disease.	1989	J Clin Invest	Dinauer MC
81	117	4.5	Cytosolic components of the respiratory burst oxidase: resolution of four components, two of which are missing in complementing types of chronic granulomatous disease.	1989	Proc Natl Acad Sci U S A	Curnutte JT
82	116	2.58	Correction of metabolic deficiencies in the leukocytes of patients with chronic granulomatous disease.	1970	J Clin Invest	Baehner RL
83	113	2.69	Chronic granulomatous disease—pieces of a cellular and molecular puzzle.	1973	Fed Proc	Karnovsky ML
84	111	22.2	Inflammasome activation in NADPH oxidase defective mononuclear phagocytes from patients with chronic granulomatous.	2010	Blood	Meissner F
85	110	15.71	Modern management of chronic granulomatous disease	2008	Br J Haematol	Segger RA
86a	109	5.19	The genetic basis of chronic granulomatous disease.	1994	Immunol Rev	Roos D
86b	109	3.41	Leukotriene production and inactivation by normal, chronic granulomatous disease and myeloperoxidase-deficient neutrophils.	1983	J Biol Chem	Henderson WR
87	108	2.45	Nitroblue-tetrazolium test for the detection of chronic granulomatous disease—technical modification.	1971	Eur J Clin Invest	Preisig E
88a	107	7.13	Long term follow-up and outcome of 39 patients with chronic granulomatous disease.	2000	J Pediatr	Liese J
88b	107	6.29	X linked chronic granulomatous disease: mutations in the CYBB gene encoding the gp91-phox component of respiratory-burst oxidase.	1998	Am J Hum Genet	Rae J

Citation classics in CGD

89	105	5.0	Long-term itraconazole prophylaxis against <i>Aspergillus</i> infections in thirty-two patients with chronic granulomatous disease.	1994	J Pediatr	Mouy R
90a	104	7.43	Clinical aspects of chronic granulomatous disease	2001	Curr Opin Hematol	Johnston RB
90b	104	3.59	DNA linkage analysis of X chromosome linked chronic granulomatous disease	1986	Proc Natl Acad Sci U S A	Baehner RL
91a	103	6.44	A novel H+ conductance in eosinophils: unique characteristics and absence in chronic granulomatous disease.	1999	J Exp Med	Banfi B
91b	103	4.68	The respiratory burst oxidase and the molecular genetics of chronic granulomatous disease.	1993	Crit Rev Clin Lab Sci	Dinauer MC.
91c	103	4.29	In vivo interferon-gamma therapy augments the in vitro ability of chronic granulomatous disease neutrophils to damage <i>Aspergillus</i> hyphae.	1991	J Infect Dis	Rex JH
92a	102	8.5	Diminished production of anti-inflammatory mediators during neutrophil apoptosis and macrophage phagocytosis in chronic granulomatous disease (CGD).	2003	J Leukoc Biol	Brown JR
92b	102	5.67	Retroviral mediated gene transfer of gp91 (phox) into bone marrow cells rescues defect in host defense against aspergillosis fumigatus in murine X linked chronic granulomatous disease.	1997	Blood	Bjorgvinsdottir H
92c	102	2.91	Mechanisms of antibody-dependent cellular cytotoxicity: the use of effector cells from chronic granulomatous disease patients as investigative probes.	1980	J Clin Invest	Katz P
93a	101	5.61	Enhanced host defense after gene transfer in the murine p47 (phox) deficient model of chronic granulomatous disease.	1997	Blood	Mardiney M
93b	101	3.61	Monoclonal antibody 7D5 raised to cytochrome b558 of human neutrophils: immunocytochemical detection of the antigen in peripheral phagocytes of normal subjects, patients with chronic granulomatous disease, and their carrier mothers.	1987	Blood	Nakamura M
94	100	9.09	Gene expression profiling provides insight into the pathophysiology of chronic granulomatous disease.	2004	J Immunol	Kobayashi SD

Citation classics in CGD

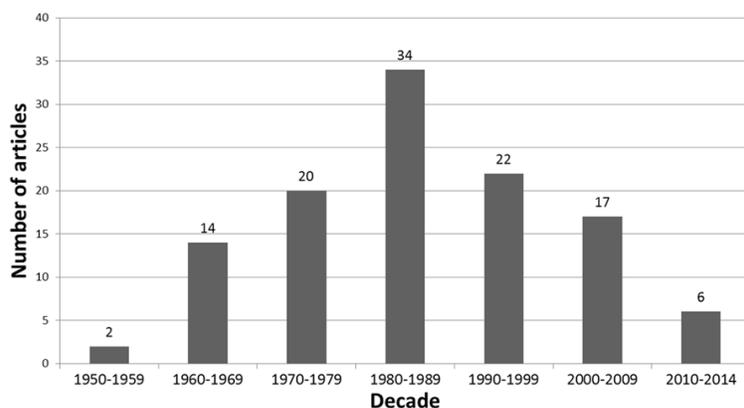


Figure 1. Number of “citation classics” articles according to decade.

infection and infiltration of viscera by pigmented lipid histiocytes”, “progressive granulomatosis”, and “granulomatosis of childhood”. The search was performed on August 28, 2016 for articles published between 1945 and the time of search. In the search, “all database” option was selected and no restrictions were used. Articles with citations equal to or exceeding 100 times were identified. The selected articles were then manually reviewed to ensure that they were related to CGD. The annual citation rates were then calculated for the selected articles by dividing the total citations number by the difference between the year of publication and the year 2015.

We analyzed the full text articles according to the total number of citations, publication year, authors, country, institution of origin, impact factor, and article type (basic science, observational study, interventional clinical trial, or review). For institution of origin, we selected the institution of the corresponding author: if authors listed more than one entity of affiliation, the largest entity such as “University” was selected; if the authors had affiliation to more than one university, all mentioned universities were selected; and if the article had more than one corresponding author, affiliations of all the corresponding authors were selected. Basic science articles included genetic studies [46], in vitro studies on human and nonhuman material, or were studies on animals, or in vivo human studies that focused on physiology or proof of principle [47]. Observational studies included case series, case-control, and cohort studies. To classify the type of the article, two study investigators (FGB and HE) reviewed all articles independently and in cases of dis-

agreement, they discussed the article until a consensus was achieved. The most recent impact factor, year 2015, was used for analysis. In cases where the journal has continued as a new title, the impact factor of the new title was used in the analysis.

Relationships between continuous variables were analyzed using Spearman’s correlation test for nonparametric data; *P* values < 0.05 were considered statistically significant. Statistical analysis was

performed by Statistical Package for the Social Sciences (SPSS) version 16.

Results

The search yielded a total of 2461 results. Of those, 115 articles were cited 100 or more times. Thus about 5% of all articles on CGD became citation classics. The articles were all in English. The oldest classic article was published in 1957 (Landing, Pediatrics) and the most recent in 2011 (Zou, Blood). The most frequently cited article received 955 citations and the least frequently cited received 100 citations (**Table 1**). The mean number of citations per article was 221.6 citations (SD = 162.7) and the median was 165 (interquartile range = 125 to 256). Twenty (17.4%) articles were cited more than 300 times. The decade from 1980 to 1989 produced the most papers with 34 articles (**Figure 1**). The most articles published within given years were 7 in year 1989 followed by the years 1987 and 1988 with 6 articles each. The annual citation rate ranged between 76.3 citations/year (Ott, Nature Medicine, at position 5) to 2.45 citations/year (Preisig, European journal of clinical investigation, at position 87). The mean number for annual citation rate was 10.1 citations/year (SD = 12.3) and the median was 7.1 citations/year (interquartile range = 4.1 to 12.4). Among the citation classics, there were 77 (67%) basic science articles, 7 (6%) interventional clinical trials, 15 (13%) observational studies, and 16 (14%) review articles.

The articles were published in 36 journals. The New England Journal of Medicine was the leading journal (16 articles, 14%) followed by Journal of Clinical Investigation (15 articles, 13%) and

Citation classics in CGD

Table 2. Journals containing the “citation classics” articles with data collected from Journal Citation Reports for the year 2015

Journal	Impact factor (2015)	Number of articles	Mean citation	Total citation	Category	Rank in category	Quartile within the category
New England Journal of Medicine	59.558	16	239.4	3831	Medicine, General & Internal	1 of 151	Q1
Journal of Clinical Investigation	12.575	15	258.3	3875	Medicine, Research & Experimental	3 of 124	Q1
Blood	11.847	11	169.5	1864	Hematology	2 of 70	Q1
Nature	38.138	6	345	2070	Multidisciplinary Sciences	1 of 63	Q1
Proceedings of The National Academy of Sciences USA	9.423	6	146.2	877	Multidisciplinary Sciences	4 of 63	Q1
Journal of Biological Chemistry	4.258	5	157.2	786	Biochemistry & Molecular biology	71 of 289	Q1
Pediatrics	5.196	5	168.8	844	Pediatrics	3 of 120	Q1
Science	34.661	5	308.6	1543	Multidisciplinary Sciences	2 of 63	Q1
Journal of Pediatrics	3.89	4	141	564	Pediatrics	6 of 120	Q1
Medicine (Baltimore)	2.133	4	417	1668	Medicine, General & Internal	40/155	Q2
Journal of Experimental Medicine	11.24	3	230	690	Immunology	7 of 151	Q1
					Medicine, Research & Experimental	4 of 124	Q1
Journal of Immunology	4.985	3	115.3	346	Immunology	32 of 150	Q1
Journal of Infectious Diseases	6.344	3	118.3	355	Immunology	20 of 150	Q1
					Infectious diseases	5 of 83	Q1
					Microbiology	14 of 123	Q1
Lancet	44.002	3	220.6	662	Medicine, General & Internal	2 of 151	Q1
American Journal of Human Genetics	10.794	2	253	506	Genetics & Heredity	8 of 165	Q1
American Journal of Medicine	5.61	2	231	462	Medicine, General & Internal	13 of 151	Q1
Journal of Leukocyte Biology (also include Journal of The Reticuloendothelial Society which Continued as Journal of Leukocyte Biology)	4.165	2	114	228	Cell biology	61 of 187	Q2
					Hematology	17 of 70	Q1
					Immunology	43 of 150	Q2
Nature Medicine	30.357	2	513	1026	Biochemistry & Molecular biology	1 of 289	Q1
					Cell biology	2 of 187	Q1
					Medicine, Research & Experimental	1 of 124	Q1
Advances in Human Genetics	NA	1	183	183	Genetics & Heredity	157 of 158	Q4
A.M.A. Journal of Diseases of Children (Continued as JAMA Pediatrics)	9.528	1	227	227	Pediatrics	1 of 120	Q1
Annals of Internal Medicine	16.44	1	206	206	Medicine, General & Internal	5 of 151	Q1
Biochimica et Biophysica Acta (continued as Biochimica et Biophysica Acta-general subjects)	5.083	1	205	205	Biochemistry & Molecular biology	52 of 289	Q1
					Biophysics	11 of 72	Q1
British Journal of Haematology	5.401	1	110	110	Hematology	10 of 70	Q1
Clinical Immunology	4.034	1	127	127	Immunology	45 of 150	Q2
Clinical Reviews in Allergy & Immunology	5.313	1	164	164	Allergy	6 of 25	Q1
					Immunology	29 of 150	Q1
Critical Reviews in Clinical Laboratory Sciences	4.167	1	103	103	Medical laboratory technology	3 of 30	Q1
Current Opinion in Hematology	3.331	1	104	104	Hematology	25 of 70	Q2
Current Opinion in Immunology	7.126	1	256	256	Immunology	15 of 150	Q1

Citation classics in CGD

European Journal of Clinical Investigation	2.687	1	108	108	Medicine, General & Internal	27 of 151	Q1
					Medicine, Research & Experimental	52 of 124	Q2
Federation Proceedings (continued as Federation of American Societies for Experimental Biology Journal (FASEB Journal))	5.299	1	113	113	Biochemistry & Molecular biology	45 of 289	Q1
					Biology	7 of 86	Q1
					Cell biology	39 of 187	Q1
Immunological Reviews	9.542	1	109	109	Immunology	8 of 150	Q1
Journal of Bacteriology	3.198	1	131	131	Microbiology	44 of 123	Q2
Journal of Laboratory and Clinical Medicine (Continued as Translational Research)	4.557	1	165	165	Medical laboratory technology	2 of 30	Q1
					Medicine, General & Internal	15 of 151	Q1
					Medicine, Research & Experimental	20 of 124	Q1
Nature Genetics	31.616	1	583	583	Genetics & Heredity	2 of 165	Q1
Pediatric Clinics of North America	2.424	1	207	207	Pediatrics	26 of 120	Q1
PLoS One	3.057	1	181	181	Multidisciplinary Sciences	11 of 63	Q1

NA: Not available.

Citation classics in CGD

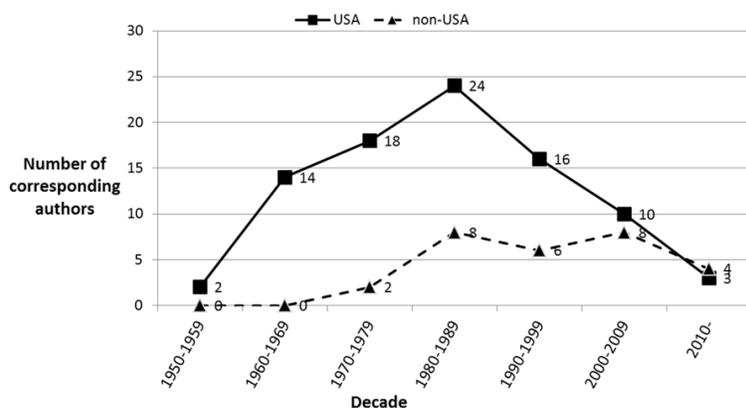


Figure 2. The number of corresponding authors broken down by USA or non-USA origin per decade.

Table 3. Name of authors with at least five “classic” articles

Author	Number of articles
Gallin JI	19
Curnutte J	18
Malech H	18
Baehner RL	13
Dinauer M	12
Holland SM	11
Orkin SH	10
Quie PG	8
Segal AW	8
Seger RA	8
Johnston RB Jr	6
Babior BM	5
Fischer A	5
Good RA	5
Holmes BM	5
Nathan DG	5
Newburger PE	5
Roos D	5
Ochs HD	5

Blood (11 articles, 9.6%) (Table 2). Five journals have continued as new titles and one publication (Advances in human genetics) had no impact factor. Of all articles, 101 (89%) were published in journals with an impact factor of more than 4. The median impact factor for the journals was 5.4 (range: 2.133 (Medicine (Baltimore))-59.5 (New England Journal of Medicine)). Journal of clinical investigation had the highest total number of citation (3875 citations). According to the ISI Web of Science Journal

Citation Report, the journals belonged to 15 categories with some journals belonging to more than one category. The most common category was “Immunology” (8 journals), followed by “General and Internal Medicine” (7 journals), and “Research and Experimental Medicine” (5 journals). Quartile order of journals within their corresponding category showed that journals were in first quartile of a category in 42 times, second quartile in 7 times, and fourth quartile in one time (Table 2).

Of the total articles, 17 (6%) were from multinational collaboration. The overall authors of the articles were from 17 countries: authors from the United States of America (USA) contributed to the highest number of articles with 91 (80%) articles, distantly followed by the United Kingdom (UK), 12 (8.6%); Switzerland, 11 (9.5%); Netherlands, 8 (7%); Germany, 6 (5%); France, 5 (4.3%); Canada, 4 (3.4%); Denmark, Italy, and Sweden, 3 each (2.6%); Hungary, Israel, Japan, and Poland, 2 each (1.7%); Belgium, Greece, and Spain, 1 each (0.8%). When only the country of the corresponding author was considered, the USA remained the most productive country with 88 (75%) articles; followed by the UK, 8; Netherland and Switzerland, 5 each; Germany, 4; France and Italy, 2 each, and Canada and Japan; 1 each. We could not determine the country of the corresponding author in one article but it could have been either from the USA or the Netherland. Analysis for publishing trends over time showed that the USA began publishing classics in 1950’s. The publication of classic articles over the years climbed and the peak was reached in 1980 to 1989. Then, USA publications began to decrease and reached a low in 2010 to 2016 (Figure 2).

A total of 465 authors contributed to the citation classics articles: Gallin JI (Director, clinical center, National Institute of Health) had the highest number, 19 articles, followed by Curnutte J, 18; and Malech H, 18 articles (Table 3). As a first author, Baehner RL had the highest number of articles (8 articles); followed by Segal AW (5 articles); Curnutte J, Dinauer M,

Citation classics in CGD

Table 4. Institution of the corresponding author with at least 3 “classic” articles

Institution of corresponding author	Number of articles
National Institute of Health (USA)	22
University of Harvard (USA)	17
University of Minnesota (USA)	8
Indiana University (USA)	7
University College of London (UK)	7
University of Washington (USA)	5
Scripps Research Institute (USA)	4
University of Amsterdam and Sanquin Research Centre (Netherland)	4
University of Zurich (Switzerland)	3

and Johnston RB (4 articles each). The mean number of authors per articles was 6.16 authors per article (SD = 6.07) and median was 5 authors (range: 1-40 authors: interquartile range: 3-7 authors). Eight articles (7%) were authored by a single author and 17 articles (15%) by 2 authors. The most common institution of the corresponding author was The National Institute of Health (NIH) with 22 articles, followed by Harvard University with 17 articles, and University of Minnesota with 8 articles (**Table 4**). The institution of the corresponding author could not be determined in five articles because the article did not identify the name of the corresponding author.

The number of authors per article had poor correlation with the number of citations per article (Spearman's coefficient 0.088; P = 0.347) (**Figure 3A**), while it had significant correlation with year of publication (Spearman's coefficient 0.521; P = 0.000) (**Figure 3B**); meaning that the number of authors per article had no effect on the number of citations an article had received and that the number of authors per article increased with time.

The journal impact factor had moderately strong positive correlation with the following: (a) the total number of citations (Spearman's coefficient 0.546; P = 0.001) (**Figure 4A**), (b) the mean number of citations (Spearman's coefficient 0.542; P = 0.001) (**Figure 4B**), and (c) the number of articles per journal (Spearman's coefficient 0.401; P = 0.017) (**Figure 4C**); meaning that the higher the impact factor of a journal, the higher the citations and the number of classics articles.

The annual citation rate correlated significantly with: (a) year of publication (Spearman's coefficient 0.695; P = 0.000) (**Figure 5A**), and (b) total number of citations (Spearman's coefficient 0.549; P = 0.000) (**Figure 5B**); meaning that as the annual citation rate increased, the year of publication and the total number of citations increased.

Discussion

We identified the list of the highest cited articles in the field of CGD. The list provides an insight into the history of research in CGD. The ranking, however, does not necessarily reflect the temporal sequence of discoveries. At position one, in 1968, Baehner and Nathan described a quantitative adaptation of the nitroblue tetrazolium test that permits precise identification of patients, carriers, and unaffected persons. By using this test and by family testing, they identified an autosomal recessive form of the disease in a girl, thus documenting a new form of inheritance for CGD [48]. The reason why this paper has been cited so often could be because the NBT test became the main diagnostic test for CGD. Moreover, the tetrazolium salts, since their introduction, have become the most widely used tools in cell biology for measuring the metabolic activity of cells [49, 50].

At position two, in 1967, Quie et al. showed that phagocytosis was normal in the neutrophils of CGD patients but the intracellular killing was defective. At position three, in 2000, Winkelstein et al., in a large retrospective study in USA, described the epidemiologic and clinical features of CGD and determined that the CGD rate of live births is 1:200,000 to 1:250,000. The oldest classic was at position 35 and was by Landing and Shirkey, in 1957. The authors, shortly after other physicians had recognized the CGD entity, had reported two boys with clinical features of CGD who had non-caseous granulomas and lipid laden macrophages in the lungs, liver, and lymph nodes. Here, a new syndrome “fatal granulomatous disease of childhood” was first described [3]. The most recent

Citation classics in CGD

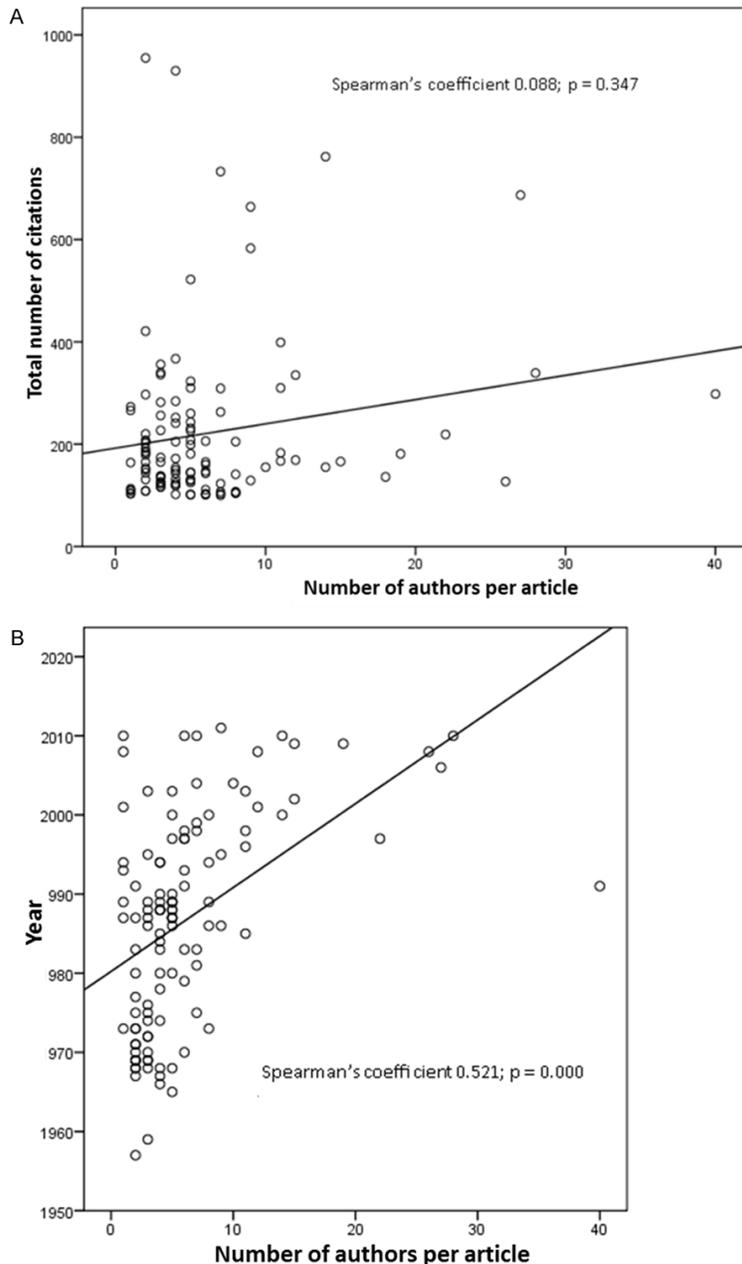


Figure 3. Scatterplot showing: A: The correlation between number of authors per article and the total number of citations per article; B: The correlation between the number of authors per article and the year of publication.

classic was at position 72 and was by Zou et al., in 2011. The authors described the generation and correction of induced pluripotent stem cells from a patient with CGD. The method represented a significant step forward to achieve reversal of the genetic defect [51].

Interestingly, the articles that had the highest and second highest annual citation rates were

related in the subject and were by the same group; Initially, Ott et al. in 2006 (annual citations rate of 76.33 and at position 5) reported on the successful use of a retroviral vector to correct the genetic defect in two CGD patients. At the time of publication, the findings were considered to have a profound implications for gene therapy [52]. However, later on, the same group published a follow up article (Stein S et al. in 2010 with 67.8 annual citations rate, at position 14) that described the adverse events and the development of myelodysplasia after gene therapy in the two boys with CGD [53].

The most active institute in production of CGD "classic" articles was the NIH with 22 articles during the period 1980-2011. Since the 1970's, the NIH clinical center has become a major site for research on CGD as it has a large CGD patient population leading to accelerate the research in treatment options such as recombinant interferon-gamma therapy, antibiotic prophylaxis, bone marrow transplantation, and gene therapy [54].

Our finding that authors from USA participated in the majority of classic articles is consistent with almost all previous studies about citation classics such as those about diabetes [8], gastric cancer [9],

gastrointestinal diseases [10], anesthesia [11], pain [12], acute pancreatitis [13], critical care medicine [7], sepsis [14], general surgery [6], obstetrics and gynecology [15], orthopedics [16], rehabilitation [17], and radiology [18]. This finding confirms the USA's overwhelming impact on medical science research because of its large population and the abundant financial resources available to the scientific community

Citation classics in CGD

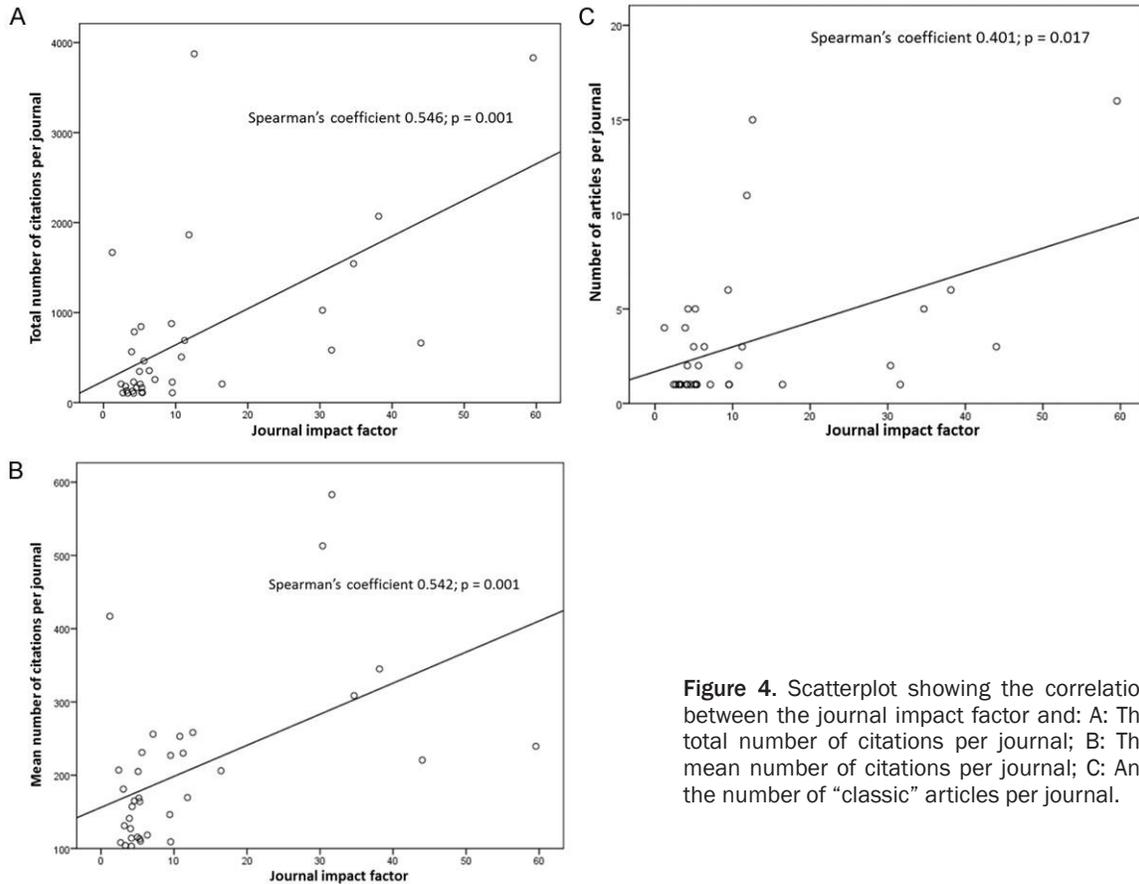


Figure 4. Scatterplot showing the correlation between the journal impact factor and: A: The total number of citations per journal; B: The mean number of citations per journal; C: And the number of “classic” articles per journal.

[12, 13, 17]. However, other possible contributing factors include: the tendency of American authors to be biased in their citation toward local articles [55] and the preference of USA reviewers to USA papers [56]. Of note, we found a trend of climbing contribution of USA corresponding authors which peaked in 1980's and then it decreased till it reached a low in 2010 to 2016. On the other hand, non-USA corresponding authors' contribution started in the 1970's and has gradually increased since then (**Figure 2**). Indeed, Stiehm and Johnston [57] have noted that although the early discoveries of CGD and the biology of the phagocytosis were made in the USA, subsequent research on phagocyte disorders and physiology has come prominently from European centers. Nevertheless, the decline of USA corresponding authors' contribution might be due to the fact that articles require certain time for recognition. Here, the most productive decade being 1980-1989 (34 articles) is consistent with the majority of reports in citation classics which suggest that a 10 to 20 years interval is

required for maximal recognition of prominent articles in a field [17]. However, our finding that the most recent article was in year 2011, and 23 (20%) articles were published after the year 2000 also indicate that CGD is a dynamic field and that many advancements have been made in recent years [27, 38, 58]. Of note, the top nine articles, according to the annual citation rate, were published between 2000 and 2011.

Here, we calculated the annual citation rate and found that it was greater for more recent articles (**Figure 5A**), a finding similar to orthopedic knee research classics [59]. This suggests that newer articles with high citation rate will have a significant impact within the next years [9]. We also found that the annual citation rate and the total number of citations showed positive correlation, contrary to orthopedic knee research classics [59].

The study has similarly identified the pioneering authors in the field of CGD. In fact, some of

Citation classics in CGD

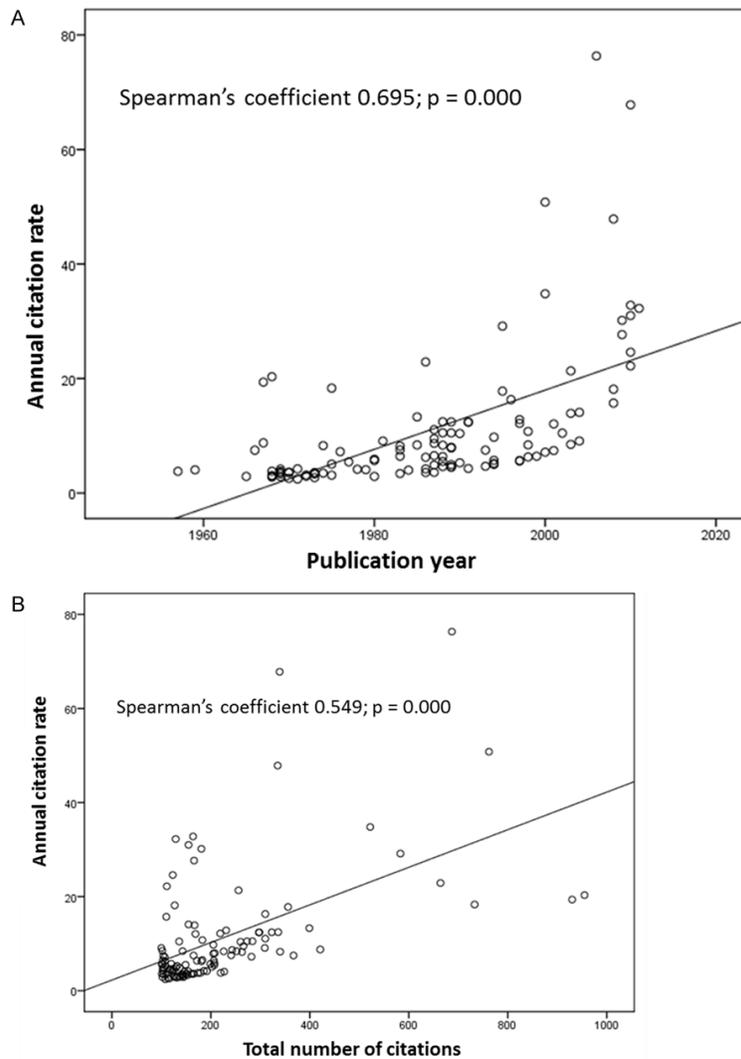


Figure 5. Scatterplot showing the correlation between the annual citation rate and: A: Publication year; B: The total number of citations.

those authors have subsequently contributed in a review or a personal account on the history of CGD research such as Robert L Baehner and Morris J Karnovsky [60], John T Curnutte [61], Robert A Good [62], Richard B Johnston [57], Steven M Holland [4], Uta Francke [63], Hans D Ochs and Walter H Hitzig [64], and John I Gallin [54].

The majority of the articles appeared in journals with high impact factor and high rank in their corresponding categories. This is consistent with previous studies on “citation classics” [17, 29-34] and is probably because the articles appeal to a large audience and are viewed as valuable by editors and reviewers [31]. We

also found that journals with higher impact factor had more citations and more “citation classics” articles. This shows the close relationship between citations and impact, and that the most cited articles are often published in journals with high impact factor, which in turn helps maintain the high impact factor of these journals [30].

We also show that the most common type of articles was of basic science similar to classics in endodontic [35] and Parkinson disease [36] and contrary to other classics such as dermatology [37], general surgery [6], obstetrics and gynecology [15], and acute pancreatitis [13]. This is probably because the rarity of CGD makes it difficult to perform clinical trials, and because the basic research in CGD has attracted more attention as it helped in understanding several mechanisms in cell biology. In addition, the wide spectrum of journals (15 categories) and the large number of authors (465 authors) reflect the great interest in CGD and the wide spectrum of its related disciplines [38].

Our study has several limitations; first, the presence of inherent problems in the citation process itself, such as incomplete or inappropriate citations, biased citation [7, 11], language bias, or self-citation [39]. Furthermore, the true milestones may in fact not be found in the most cited articles themselves but in their reference lists, underlining the fact that the number of citations an article has accumulated cannot be used as a sole measurement of its importance [11]. Second, we restricted the search to titles with the term “chronic granulomatous disease”, or one of its other names. However, while this method was used by others [32, 36, 40], this could have resulted in missing relevant articles [32, 36]. An example of such missed articles is

the one by Holmes B et al. "Studies of the metabolic activity of leukocytes from patients with a genetic abnormality of phagocytic function" (658 citations). Nonetheless, we used this strategy because it is reproducible and it avoids the subjective bias in deciding the relevance of a particular article. Third, we used the cutoff of 100 citations; important work might have been missed because either the work is recent and did not accumulate enough citations or the work was cited few times until its findings became well known, a phenomenon called "obliteration by incorporation" [15]. To partially control for this bias, we calculated the annual citation rate for the included articles. However, this would control only for articles ranking within the list but would not rank articles with high annual citation rate if they have total citations of less than 100. Fourth, we used a single citation database; the ISI Web of Science. Other available databases include Google scholar and Scopus. In ISI web of Science, only a select set of journals are searched. Moreover, ISI web of Science does not allow for analysis of a large number of foreign articles because it uses only English citations. However, we opted to use this database because it is the most commonly used and is a robust method for clinical medicine [6, 15-17, 25, 36], and because it yields similar results to Google scholar and Scopus in small fields [34, 41]. Fifth, we used the institutions of the corresponding author in the analysis, thus we might have missed some institutions. Sixth, we relied on journal impact factor for the year 2015; articles published long ago might have originally been associated with a different impact factor since the impact factor of a given journal varies with time [32]. Seventh and finally, the list of citation classics changes with time and therefore is a snapshot of the current state of research [41]. However, despite these limitations, our study provides some insights into the most cited research articles in CGD since its first description 60 years ago.

In conclusion, despite the above limitations, our study provides some insights into the most cited research articles in CGD since its first description 60 years ago. In addition, the study shows that most of the "Citation classics" in CGD were from USA, were published in high impact factor journals, and were of basic science type. Furthermore, the CGD research is a dynamic field and has a wide spectrum of interest across several medical disciplines.

Disclosure of conflict of interest

None.

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