HHS Public Access

Author manuscript

Clin Immunol. Author manuscript; available in PMC 2021 April 25.

Published in final edited form as:

Clin Immunol. 2010 June ; 135(3): 347–363. doi:10.1016/j.clim.2010.01.006.

Good's syndrome remains a mystery after 55 years: A systematic review of the scientific evidence

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Abstract

Good syndrome (GS) is a rare association of thymoma and immunodeficiency first described more than 50 years ago. However, this syndrome still remains a mystery to clinicians. We systematically reviewed all the clinical, laboratory and immunologic findings from 152 patients with Good syndrome. The syndrome has a worldwide distribution and approximately half of the cases (47%) have been described in Europe. The diagnosis of thymoma preceded the diagnosis of hypogammaglobulinemia, infection, or diarrhea in 42% of patients whereas in 38% of patients the diagnoses were made almost simultaneously within 2 months of each other. We found significant mortality in patients with this syndrome (44.5%). Astute clinical acumen and increased awareness about the clinical and immunological profile of this syndrome may increase early recognition of this syndrome and prevent mortality. Further studies are needed to elucidate this clinical entity.

Keywords

Good's syndrome; Immunodeficiency; Infections; Hypogammaglobulinemia; Thymoma

Introduction

Good syndrome (GS) is a rare association of thymoma and immunodeficiency first described more than 50 years ago. Patients are most commonly between the ages of 40 and 70 years and have a thymoma, low to absent B cells in the peripheral blood, hypogammaglobulinemia, and defects in cell-mediated immunity. Although this syndrome has been reviewed previously [1,2], the only published systematic analysis of the infectious complications associated with GS was based only on 46 cases and a systematic analysis of all the clinical, laboratory and immunologic findings was not performed. Numerous reports on GS have been published since that report and this syndrome still remains a mystery to

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Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.clim.2010.01.006.

clinicians. Thus, we reviewed the literature and systematically summarize the information from 152 patients with GS.

Methods

All previous cases included in our literature review were found using a Pubmed search (1956-December 2009) of the medical literature applying the terms “thymoma,” “hypogammaglobulinaemia,” “immunodeficiency,” “Good’s syndrome,” and “infection.” Additional cases were identified using references of publications found. Only articles and abstracts published in English were included in our final analysis.

Definition

The association between the presence of a thymoma and adult-onset hypogammaglobulinemia was first described by Dr Good in 1955. There are a number of definitions for Good syndrome. Many authors choose to define it as hypogammaglobulinemia with thymoma consistent with Dr. Good’s case. Practice parameters in 2005 [3] define it as a subset of common variable immunodeficiency (CVID), however, the reduced numbers of peripheral B cells noted in Good syndrome are not a feature of common variable immunodeficiency, which typically shows impaired B-cell maturation. Although there are no formal diagnostic criteria for this disorder, it is classified as a distinct entity by the expert committee of the World Health Organization and International Union of Immunological Societies on primary immunodeficiencies [4]. Since the pathogenesis of the disease remains unknown and patients have several other immunological impairments in addition to hypogammaglobulinemia, Good syndrome may better be defined as immunodeficiency with thymoma, a broader classification. Hence, it is described as a constellation of thymoma and adult-onset immunodeficiency characterized by hypogammaglobulinemia, low or absent B cells, variable defects in cell-mediated immunity with a CD4 T lymphopenia, an inverted CD4/CD8+ T-cell ratio and reduced T-cell mitogen proliferative responses.

Results

Selected studies

We used the simultaneous presence of thymoma and hypogammaglobulinemia as the minimal diagnostic criteria for a diagnosis of Good Syndrome. Using the key terms, 241 patients with the syndrome were reported in 188 studies [1,2,5–190], 113 (46.9%, 113/241) patients were reported in Europe [1,5,7–9,11,12,15,16,18,21–25,28,31,32,34,35,38,39, 43,46,47,49,50,53,54,57,58,120,124–128,135,136,140,179, 181,184,185], 71 (29.5%, 71/241) in America [2,27,29,36,44, 45,70,74,85,95,99,105,110,113,116,122,132,134,137–139,142,146,147,149,150,152,154,156,159,166–169,172–176,180,182,183,187–190], 55 (22.8%, 55/241) in Asia [6,10, 13,14,17,19,20,26,30,33,37,40–42,48], 1 (0.4%,1/241) in Africa [93], 1 (0.4%,1/241) in Oceania [76]. 134 of these studies were from the English literature [1,2,5–10,13–23,25–32,35–42,44–46,51–54,56–60,64,65,69,70,73–78,81–83,85–88,93–99,101,102 – 105,108,110,112 – 117,119, 122–124,127,128,132–142,144–150,152,154–157,159, 166–169,171–176,179,180–190]. Seven studies [53,88,96,

97,137,139,146,152] did not include detailed data about the patients and were not included in our final analysis while 54 studies were from the non- English literature and were also excluded [11,12,16,24,33,34,43,47–50,55,61–63,66–68, 71,72,79,80,84,89–92,100,106,107,109,111,118,120,121, 125,126,129–131,143,151,153,158,160–165,170,177,178]. Thus, we included 127 studies in our final analysis which described 152 patients (Appendix). [1,2,13,17,21,22,23,25–27, 29,30–32,35–38,41,42,51,56,58,59,65,69,71,74,75, 77,81,83,85,93,98,101,102,103,105,113,114,116,122,123, 133,137,138,141,148,155,157,159,166,167,168,171,172,175, 182,183,187–189,191–208]. However, the vast majority of these publications are isolated case reports or enumeration of patients with coexisting hypogammaglobulinemia among patient series with thymoma, without further descriptions or follow-up details. To our knowledge, there are only 10 reports in the English literature describing details from case series with more than 2 patients with Good's syndrome, 2 case series with eight patients [46,152], one with seven [88], one with 5 patients [2], and 6 with 3 patients [1,96,103,136,146,180]. Nevertheless, only few of them reported detailed data about these patients [1,2].

Patients

127 cases reported in the literature met our criteria for inclusion (see Methods) and a total of 152 patients with GS were analyzed in this review. There were 79 female patients and 73 males. With the exception of a single pediatric case [99], the mean age of the patients was 59.1 years (range, 25–90 years, SD 12.1) at the time of the first manifestation (thymoma, infection or hypogammaglobulinemia). There was no statistical significant difference between the mean age at the time of the first manifestation of GS in men (58.0 ±12.4) vs. women (60.3±11.8). In 20 patients the initial presenting symptom and sequence of diagnoses (thymoma, infection, and immunodeficiency) was not specified [23,46,103,108,122,136,180]. Thus we extrapolated data regarding initial presentation of GS in 132 patients with available data.

Clinical manifestations of GS

Thymoma—The diagnosis of thymoma preceded the diagnosis of hypogammaglobulinemia, infection, or diarrhea in 56 (42.4%, 56/132) patients [1,2,5,7–10,14,19,20,26–28,36,37,45,51, 52,54,58,69,74,78,83,86,98,99,101,105,110,113,119,135, 138,145,147–149,154,155,157,159,167–169,173–175,181, 183,186–188] with an interval of 3 months to 18 years. Thymoma was diagnosed after the documentation of infection or hypogammaglobulinemia in 26 (19.7%, 26/132) patients [1,2,29,35,38,39,57,70,93–95,102,104,114,123,128,134,144,156,179,182,185,189,190], with an interval of 3 months to 15 years. In 50 (37.9%, 50/132) patients, the diagnoses were made almost simultaneously within 2 months of each other [1,2,6,13,15,17,18,21,22,25,30–32,40–42,44,56,59,60,64,65,73,75–77,81,82,85,87,112,115–117,124,127,132,133,140–142,150,166,172,176,184,186], and in 8 patients the thymoma was only diagnosed at autopsy [1,103,119,123,141,174,187,190].

The histologic types of thymomas identified in patients with GS are described in Table 1.

Infections—The infectious diseases and the recovered pathogens from patients with GS are shown in Table 2. In 24 patients (16.7%) with this immunodeficiency no infections were reported [32,81,82,103,108,110,115,116,122,124,136,141,156,157,169,179,180].

Gastrointestinal manifestations—Diarrhea was present in 31.8 % of patients (42/132 of patients with available detailed data on clinical presentation) [1,2,8,18,26–28,37,40–42,59,64,70,74,82,86, 105,112,113,117,123,128,133,134,138,140,142,150,155,157,159,167,172–174,181,184,186,188,189]. The mechanism of diarrhea was not identified in the majority of cases. Infection was the etiology of diarrhea in 15 patients (35.7% of 42 cases) [2,18,27,41,42,59,64,86,102,112,113,117,128]. Seven patients had bacterial diarrhea [2,18,27,64,86,112] and *Salmonella* spp was the most common pathogen (50%, Table 1) [2,27,64,86,112]. *Giardia lamblia* and Cytomegalovirus (CMV) were the cause of diarrhea in four [41,102,113,128] and five [2,42,59,64,117] cases respectively. Two patients had CMV colitis and bacteremia secondary to *Campylobacter jejuni* [2,42]. Ulcerative colitis as a cause of diarrhea was described in 2 cases of Good's syndrome [12,167], while immune mediated colitis as cause of diarrhea was also suggested in 2 more cases [1]. Primary sclerosing cholangitis has also been described in one patient with this syndrome [82].

Autoimmune manifestations—We identified 89 patients (58.6% of 152 patients included in our review, Table 3) with autoimmune manifestations, such as myasthenia gravis, pure red cell aplasia (PRCA) and other hematological abnormalities [6,9,19,22,32,40,60,78,82,83,102,110,114,117,136,144,157].

Laboratory findings

The laboratory findings in patients with GS are presented in Table 4. The HIV test was negative in all the cases of GS that reported this test [2,5–10,13,14,19,22,179].

Outcome

From 152 patients included in our review data on the outcome were available for 128 patients. No data on the outcome were reported in 24 patients [1,27,31,35,41,46,65,77,82,94,95,104,122,134,136,147,149,183,210]. For 48 patients (37.5%) there was improvement in the outcome (defined as reduction in the incidence of infections) after treatment [1,5,13,15,17,20,22,25–27,29,30,36,39,42,51, 52,54,58,60,64,65,69,70,73,76,78,83,85,87,98,102,103,112,114,115,124,127,128,133,144,154,167,189,212] whereas for 23 patients (18.0%) there was no difference in the outcome after treatment [6,8–10,18,21,32,54,56,57,74,81,86,116,132,134,148,155,169,179,181,182]. 57 patients (44.5%) died during the observation periods included in the case reports. 34 patients (59.6%) died as a result of an infection [2,7,14,19,23,28,37,40,44–46,59,75,93,99,101,115,117,138,140,142,150,156,157,159,166,172,175,185,186], 15 patients (26.3%) died as a result of a known non-infectious cause [2,38,105,110,135,145,168,171,173, 175,176,184,186,188] and in 8 cases (14.1%) the cause of death was unclear [1,103,119,123,141,174,187,190]. Nonsurgical regression of thymoma following immunosuppressive therapy was noticed in one case [64].

Discussion

Fifty-five years after Dr. Good's original observations on thymoma and hypogammaglobulinemia, the pathogenesis of this primary immunodeficiency syndrome remains incompletely understood. We performed a systematic review of the literature on this clinical entity based on data from 152 cases.

Epidemiology

The syndrome has a worldwide distribution and most cases have been described in Europe. In the United States, the incidence of thymoma is 0.15 cases per 100,000 [213], and this tumor can be found in up to 10% of patients with adult-onset hypogammaglobulinemia [214]. On the other hand, the incidence of hypogammaglobulinemia is 6–11% in patients with thymoma [214,215]. In one study, which was probably influenced by referral bias, Good's syndrome was noted in 7% of adults with primary antibody deficiency attending a chest clinic [139]. This syndrome is the underlying cause of immunodeficiency in 1% to 2% of patients with primary antibody deficiency who are on immunoglobulin replacement treatments [4]. In contrast to most other primary immunodeficiency states, GS usually presents in the fourth or fifth decade of life, although it can very rarely also occur in children. Similarly to previous reports [195], we found that male and female patients are equally affected in GS and that the mean age at presentation was 59.1 years while only one case was diagnosed in childhood [99].

Pathogenesis

The pathogenesis of the immunodeficiency in GS remains elusive. The principal immunological findings in Good's syndrome are hypogammaglobulinemia, few or absent B cells, abnormal CD4+:CD8+ T cell ratio, CD4 T cell lymphopenia, and impaired T cell mitogenic response. Almost all patients had reduced levels of immunoglobulins. A reduced mature B cell count or even the absence of B cells from peripheral blood was noted in 87% of cases. Such flow cytometric findings in the peripheral blood do not imply total disappearance of B cells. B cells might exist in the spleen, lymph nodes and other lymph tissues, assuring immunoglobulin productions, although significantly inhibited. The presence of B and T cell lymphopenia, pre-B cell arrest, impaired maturation of erythroid and myeloid precursors, pure red cell aplasia, neutropenia and eosinopenia in many cases of GS may suggest that the basic defect may be in the bone marrow. The absence of pre-B cells has also been reported in bone marrow samples from patients with GS [212]. Lymphocytes from bone marrow were abnormal in 17 patients with GS [8,18,19,38,42,51,56,64,69,77,87,112,113,115,134] and were normal in 11 patients with Good's syndrome [2,21,30,52,60,98,116, 128,155,172,183]. Thus, the pathogenesis of GS is quite heterogeneous especially since T cells abnormalities have also been found in thymic tissue in 3 cases [112,116,122]. Interestingly, plasma cells were absent from lymph nodes, gut associated lymphoid tissue and spleen in six [112,168, 171,173,182,190],2 [112,173] and 2 cases [171,173] respectively, suggesting that lymphocyte abnormalities may be more diffuse in patients with GS and may not be limited in bone marrow. In four cases of PRCA, the authors identified increased (rather than reduced) number of lymphocytes, including T cells, in the bone marrow [69,113,115], suggesting that the population of lymphocytes in the

bone marrow may be different in cases of GS with presence of autoimmune phenomena such as PRCA compared to other cases. The presence of autoantibodies in many cases may also suggest autoimmune pathogenesis. However, the immunologic abnormalities do not correct following either corticosteroid treatment or thymectomy.

T cell defects can be manifested by cutaneous anergy to 2 or more test antigens (such as tuberculin) and impaired response to mitogens suggests absent delayed hypersensitivity. We found that > 60% of patients with GS can have these abnormalities. Decreased *in vitro* IL-2 production in response to stimulation was noted in the T cells from 2 patients that were tested [2,74], which may suggest that patients with Good's syndrome have activated memory T cells that have dysregulated cytokine production. In one study the percentage of naïve CD4 and CD8 cells was reduced in bone marrow but the percentage of memory T cells and the total number of T cells in bone marrow were normal [8].

The HLA subtype was determined in only 3 studies and was A2, B27, and BW4 in one study [70] and A3, 29, B51,DR4 [83] and A2,3, B7,15, Cw3, DR2 [114] in the other 2 studies. HLA A2 [70,114] and A3 [83,114] was found in 2 studies. Although functional abnormalities of white blood cells have not been described as part of GS we found 2 studies that reported abnormal chemotaxis of polymorphonuclear cells [93,182]. In addition, leukopenia and neutropenia were present in 46% and 15% of cases with GS. Nonetheless, the wide range of normal and abnormal immunologic values reported in the literature suggests individual variations in the nature or severity of this immunodeficiency. Thus, further studies are needed to elucidate the complex immune abnormalities that are present in patients with GS.

Clinical manifestations

Patients with GS usually develop either thymoma or infectious complications of their immunodeficiency, with or without the development of autoimmune manifestations, usually within approximately 6 years of the first presentation [216].

Thymomas

We found that the diagnosis of thymoma may precede the diagnosis of hypogammaglobulinemia, infection, or diarrhea by 3 months–18 years in 42% of patients with GS. Patients may complain of symptoms secondary to the thymoma itself such as cough, dysphagia, and hoarseness. Thymomas can be a subtle feature on chest *x* rays, and in one study 25% of tumors were missed, with a diagnostic delay of 41 months [217]. A computed tomography (CT) scan or magnetic resonance imaging of the chest have increased sensitivity for detection of thymomas and can also define the clinical stage of this tumor [218,219]. We found that the most common histologic type of thymoma in GS was a spindle cell variant according to the traditional classification of thymomas and the AB variant according to WHO classification of thymomas. Malignant thymic carcinomas were identified in <10% of cases.

Infections

Patients with GS have increased susceptibility to bacterial, fungal, viral, and opportunistic infections related to both humoral and cell-mediated immune deficiencies (Table 2) [2,195]. We found that recurrent infection of the upper and lower respiratory tract was the most common infection reported. *Haemophilus influenzae* and *Pseudomonas* spp were the most common pathogens identified. CMV and *Candida* spp were the most common opportunistic pathogens reported. The occurrence of opportunistic infections in GS suggests that patients have severe defects in cell-mediated immunity. However, in contrast to human immunodeficiency syndrome, there were only two cases of *Mycobacterium tuberculosis* and one case of *Toxoplasma gondii*. No *Cryptococcus neoformans* infection has been described in Good's syndrome. Whether the prevalence of these organisms in patients with GS is different to that documented in other patients with disorders of cell mediated immunity is not known.

Diarrhea

Diarrhea is present in almost 50% of patients with Good's syndrome, and in most cases it is chronic [2]. It has rarely been associated with malabsorption due to villous atrophy [117]. Although enteric bacteria (mainly *Salmonella* spp), *Giardia lamblia* and CMV were occasionally isolated, in the vast majority of cases we did not identify definite pathogens [212]. The presence of inflammatory lesions similar to those seen in inflammatory bowel disease and the response to treatment with local anti-inflammatory agents and systemic steroids may suggest an autoimmune basis of the colitis in some cases [167]. However, since there are no systematic studies evaluating the nature of idiopathic diarrhea in this disorder, the cause of the inflammatory diarrhea associated with Good's syndrome remains unknown.

Autoimmune manifestations

We found that the most common autoimmune syndrome associated with GS was PRCA followed by myasthenia gravis. Previous reports have estimated that autoantibodies can be present in up to 30% of cases of GS, despite low immunoglobulin synthesis [166]. We found that the most common autoimmune antibodies were antinuclear antibodies (ANA) which were present in more than half the cases of GS in which autoantibodies were measured. Although ANA can be found in up to 3% of patients with Good's syndrome [166], in most cases autoantibodies were either not measured or were measured soon after recovery from a severe infection. Thus, the significance of presence of autoantibodies in a state of immunodeficiency remains unclear but their presence may suggest a role in the pathogenesis of hypogammaglobulinemia.

Laboratory findings

The laboratory findings of Good's syndrome are summarized in Table 4. Physicians should have a high level of suspicion for diagnosis of certain infections since serologies may be unreliable in this state of immunodeficiency and detection of antigens using PCR should be undertaken.

Prognosis

The prognosis in patients with Good's syndrome is believed to be worse than in those with other immunodeficiencies [2]. The possibly worse clinical outcome of patients with GS, compared with patients with CVID, appears to be determined by the severity of associated infectious, hematologic, and autoimmune diseases [220], rather than by the behavior of the thymoma. In addition, the clinical course of disease may be more severe for those patients who require immunosuppressive agents for associated autoimmune processes. In a single centre review of primary antibody deficiency spanning 20 years, 70% of patients with Good's syndrome were alive 5 years after diagnosis compared with almost 100% of patients with CVID [88]. At 10 years, only 33% were alive compared with 95% of patients with CVID. Finally, we also found significant overall mortality (46%) in our review of 152 cases of GS.

Management of Good's syndrome

Thymectomy—The treatment of thymoma is surgical removal or debulking of the tumor, [216,219,220] and the most important indicator of long term prognosis is completeness of tumor resection [219,221]. Patients with advanced stage 3 or stage 4 disease tumors often require radiotherapy and combination chemotherapy. Thymectomy can prevent locally invasive growth and metastasis of thymomas and usually has a favorable effect on associated conditions like myasthenia gravis and pure red cell aplasia but does not usually reverse the immunological abnormalities [215].

Gamma globulin—Immunoglobulin replacement treatment has been reported to improve infection control, reduce hospitalization, and decrease use of antibiotics. The more effective intravenous immune globulin (IVIG) was used since 1979. Poorer responses were seen with the intramuscular preparation (IMIG) that was used in early reports. A retrospective review of the efficacy of immunoglobulin treatment in this disorder showed that 23 of 30 patients had a reduction in the numbers of bacterial sinopulmonary infections [2]. In our review, approximately 38% of patients with GS had reduced incidence of infections after treatment with gamma globulin. Thus, IVIG is recommended as a means of maintaining appropriate immunoglobulin G levels for all these patients.

Other therapeutic measures—Other treatment modalities have also been tried such as immunosuppressive therapy, plasmapheresis [128], splenectomy [157,169,186,187], transfer factor from human leukocytes [133], but all had moderate results.

Preventive measures—Immunological investigations, including quantitative immunoglobulins, B cells, T-cell subsets, should be considered part of the routine diagnostic evaluation in patients with thymoma and infections. If immunologic test results are normal, testing should be performed periodically if the clinical suspicion of GS persists, because there can be an interval between the diagnosis of immunodeficiency and/or thymoma and development of infection [212].

IgG antibodies to toxoplasmosis and cytomegalovirus should be determined to evaluate whether a patient is at risk of reactivated infections. Patients with Good's syndrome who are

CMV antibody negative or whose CMV serology is unknown, or cannot be determined, should receive CMV negative blood to avoid the potential risk of iatrogenic disease [59]. Graft versus host disease is a complication of malignant thymoma and it would be prudent to use irradiated blood in patients with Good's syndrome [197,222]. In one study of yellow fever immunization, 4 out of 23 vaccinees that developed vaccine associated viscerotropic disease had an underlying thymoma (and presumably Good's syndrome) [223]. Thus, because of the T-cell defect, the use of live vaccines in patients with Good's syndrome poses a significant risk and should be avoided. Appropriate microbiological investigations and prophylactic antibiotics are warranted. In certain cases of GS co-trimoxazole according to the regimen recommended for AIDS patients with CD4 cell count less than 200 cells/mm^3 was given [20].

Limitations

This review has certain limitations. We included publications written only in the English language for our final analysis. Unfortunately in most cases there was incomplete microbiologic or immunologic evaluation, documentation of clinical findings and lack of long-term follow-up. GS is most likely underreported since it may go unnoticed when thymoma is not considered clinically (given its rarity). Most patients' immune status was evaluated on a single occasion, and many reports date back to the pre-flow cytometry era before lymphocyte subsets could be studied. Immunologic data were available on fewer than half of the patients and need to be interpreted with caution in the absence of controlled studies.

Conclusions

Good's syndrome should be considered in patients over 40 years of age with unexplained antibody deficiency. In patients with a thymoma, recognition of hypogammaglobulinemia is an important clinical consideration, since GS has significant mortality. The major causes of death in patients with GS include infections, autoimmune diseases, and hematologic complications. IVIG can reduce the risk of infections, excess antibiotic administration and hospitalizations and should be given if humoral immunodeficiency is demonstrated. Astute clinical acumen and increased awareness about the clinical and immunological profile of this syndrome, may increase early recognition of this syndrome and prevent mortality. Further studies are needed to elucidate this clinical entity.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Table 1

Histologic type of thymomas in 152 patients with Good syndrome according to the traditional and WHO classification of thymomas.

Histologic type of thymoma	Number of cases (%)	References
Traditional classification of thymomas	100	[1,2,7,14,15,28,36,38,39,42,45,54,65,75,81,85,93,98,101,114,115,124,133,134,139,145,148,168,169,171,175,183–186,190]
Spindle cell thymomas	52/100 (52%)	[44,52,64,78,88,117,119,134,139,154,156,157,176]
Lymphoepithelial tumor	19/100 (19%)	[1,2,14,15,39,42,98,114,115,124,133,134,148,168,169,184,185]
Epithelial thymoma	11/100 (11%)	[28,65,101,108,145,171,183,186,190]
Malignant thymoma	10/100 (10%)	[36,38,45,54,81,88,93,96,186]
Lymphocytic type	3/100 (3%)	[115,139,175]
Mixed cell type medullary type	2/100 (2%)	[35,60]
Medullary thymoma	2/100 (2%)	[1,75]
Lipothymoma	1/100 (1%)	[85]
WHO classification of thymomas	24	[5,6,8–10,13,17–20,26,30–32,38,40,46,181]
Type AB	10/24 (41.7%)	[5–9,13,17,20,32,46]
Type B2	6/24 (25%)	[19,30,38,46]
Type B1	3/24 (12.5%)	[26,46]
Carcinoma	2/24 (8.3%)	[46]
Type A	2/24 (8.3%)	[18]
Type B3	1/24 (4.2%)	[31]
Histologic type not specified	28	[21,23,25,27,29,37,41,54,57,58,70,74,77,82,83,86,87,94,95,99,104,105,110,128,135,136,138,141,142,155,159,167,172–174,179–182,189]

Table 2

Infections described in patients with Good syndrome.

Infection	No. of patients	Pathogens (*n*, %)	References
Bacterial infections			
Recurrent sinopulmonary infection	98 (30 had available culture data and 53 isolates were identified)	*Haemophilus influenzae* [a] (13, 24.5%), *Pseudomonas* spp. (12, 22.6%), *Klebsiella* spp. (7, 13.2%), *Streptococcus pneumoniae* (7, 13.2%), *Staphylococcus aureus* (4, 7.5%), *Moraxella catarrhalis* (2, 3.8%), *Serratia* sp (2, 3.8%), *Streptococcus viridans* group B (1, 1.9%), *Salmonella* sp (1, 1.9%), *Enterobacter* sp (1, 1.9%), *Acinetobacter baumannii* (1, 1.9%), *Mycoplasma pneumoniae* (1, 1.91%) *Candida albicans* (1, 1.91%) A pathogen was not isolated in 68 cases of recurrent sinusitis, bronchitis, or pneumonitis which were documented to be considered of possible infectious pathogenesis.	[1,2,5,7–10,14,15,18,19,21,25,26,27,29,31,35,37,39–42,44–46,51,52,54,57,59,65,69,73–76,78,83,85,93–95,101,104,112,114,119,123,124,128,132,133,134,138,142,145,147,148,150,154,155,157,159,166–168,171–176,181–187,189,209]
Other lung infections	18	Necrotizing pneumonia with lung abscess (1), mediastinitis (1), empyema (1), bronchiectasis (15)	[2,8–10,19,54,95,134,142, 147,148,154,175,176,181,182,184]
Bacteremia	16 (13 had available culture data	No pathogen recorded (3) *Campylobacter spp* (5, 38.5%), *H. influenzae* (1, 7.7%), *Klebsiella* spp. (1, 7.7%), *Pseudomonas* spp. (1, 7.7%), group A streptococcus (1, 7.7%), *Alcaligenes xylosoxidans* (1, 7.7%), *Salmonella* spp (1, 7.7%), *E. coli* (1, 7.7%), Gram-negative bacteria (1, 7.7%)	[2,5,23,42,56,60,64,87,140,150,187,190]
Bacterial diarrhea	7 (6 with culture data)	*Salmonella* spp. (3), *C. jejuni* (2), *Clostridium difficile* colitis (1), no pathogen recorded (1)	[2,18,27,42,64,86,112]
Urinary tract infections	5 (2 cases with culture data)	*Proteus* spp. (1), *E. coli* (1)	[159,172–174,189]
Skin infections	2	Furunculosis and skin abscess (1), wound infection, (*C. perfringens*) (1)	[157,188]
Bone and joint infections	2	Osteomyelitis (*Alcaligenes xylosoxidans)*, arthritis *Mycoplasma* spp. (1)	[23,86]
Mycobacterial infections			
Tuberculosis	2	Pulmonary (1), pulmonary and otic (1)	[114,174]
Other mycobacteria	1	Bronchopulmonary infection with *Mycobacterium malmoense*	[77]
Viral infections			

Infection	No. of patients	Pathogens (*n*, %)	References
CMV disease	25	Retinitis (8), pneumonia (5), encephalitis (3), colitis (3), enteritis (3), adrenalitis (2), gastritis (1),	[2,5,12,20,28,36,42,44,54,58,59,64,65,113,117,138,150,175]
CMV infection of uncertain clinical significance	7	CMV infection of tumor cells (1), CMV serology without any other symptoms (1) Asymptomatic viruria (2), CMV recovered from sputum (2), CMV inclusions at autopsy in: salivary glands (1), thyroid (1), parathyroid (1), lungs (7), trachea (1), Kaposi cells (1), esophagus (1), pancreas (1) prostate (1), spleen, adrenals (2), liver (1), colon (1).	[101,113,142,150,173,175,182]
Varicella-zoster virus	11	Disseminated varicella (1), herpes zoster (10)	[20,37,64,65,75,99,113,127,144,182,189]
Herpes simplex virus (HSV)	8	Skin infections (4) Epiglottitis (1), tracheobronchitis (1) keratitis (1), recurrent genital herpes (1), ulcerative dermatitis (1, possibly related to HSV), respiratory infection (2) Disseminated HSV infection (1)	[11,16, 93,94,98,117,135,145]
HHV-8	3	Kaposi sarcoma	[27,75,101]
HPV	1	Epidermodysplasia verruciformis	[93]
Echovirus 22	1	Cultured from ascitic fluid	[2]
JC virus	1	Progressive multifocal leucoencephalopathy	[7]
Fungal infections			
Candida infections	29	Persistent mucocutaneous candidiasis (5), oropharyngeal candidiasis (10), esophagitis (6), candidemia (3), lung infection (2), oral–intestinal candidiasis at autopsy (1), candidal pyelonephritis(1), unspecified Candida infection (5)	[1,2,14,15,22,28,37,38,41,42,44,45,59,60,64,73,75,83,96,101,105,113,117,128,133,150,186,189]
Pneumocystis jirovecii (carinii)	9	Pneumonia	[2,14,36,40,59,76,96,98,173]
Aspergillus	4	Aspergillosis (1), isolation of Aspergillus from BAL and sputum (2), Disseminated aspergillosis (1)	[1,38,96,157]
Parasitic infections			
Giardia lamblia	4	Giardiasis	[41,102,113,128]
Babesia microtii	1	Babesiosis	[27]
Toxoplasma	1	Ocular toxoplasmosis	[126]
Other infections			
Recurrent eye infections such as in 3 patients,	3	Recurrent scleritis, cicatrizing keratoconjuctivitis conjunctivitis	[30,112,172]
Genital infections	2	Orchitis (1), penile infections (1)	[45,172]

Infection	No. of patients	Pathogens (*n*, %)	References
Lymphadenitis	1	Cervical lymph nodes (1)	[189]

[a]Numbers in parenthesis represent % of total isolates (48 bacterial isolates for respiratory infections).

Table 3

Autoimmune manifestations described in 89 patients with Good syndrome.

Autoimmune manifestation	Number of patients (%)	References
Pure red cell aplasia (PRCA)	31 (34.8%)	[1,13,17,25,65,69,85,96,103,113,115,116,119,122,145,150,169,171,175,176,180,187,188]
Myasthenia gravis	14 (15.7%)	[32,102,114,117,134]
Oral lichen planus	11 (12.4%)	[9,21,22,27,29,30,59,73,85,105,148]
Aplastic anemia	7 (7.9%)	[141,147,156,166,168,180,185]
Macrocytic anaemia	5 (5.6%)	[60,83,110,114,157]
Autoimmune hemolytic anemia	3 (3.4%)	[175,187,210]
Monoclonal gammopathy	3 (3.4%)	[19,78,83]
Myelodysplastic syndrome	2 (2.2%)	[15,42]
Diabetes mellitus	2 (2.2%)	[78,114]
Polyarthropathy	2 (2.2%)	[136,144]
Paroxysmal nocturnal hemoglobinuria	1 (1.1%)	[32]
Agranulocytosis	1 (1.1%)	[166]
Thrombocytopenia	1 (1.1%)	[60]
Idiopathic myelofibrosis	1 (1.1%)	[78]
Dermatomyositis	1 (1.1%)	[123]
Primary sclerosing cholangiitis	1 (1.1%)	[82]
Sweet’s syndrome	1 (1.1%)	[78]
Ulcerative colitis	1 (1.1%)	[167]
Vulvovaginal gingival lichen planus	1 (1.1%)	[22]

Table 4

Laboratory findings in patients with Good syndrome with available data.

Laboratory data	Number of patients with available data (%)	Comments	References
Anemia	74/87 (85%)	13 patients had normal hemoglobin. PRCA was the mechanism of anemia in 31 (41.9%) patients	[17,18,32,39,41,42,52,60,64,65,69,73,78,83,93,95,110,115,117,119,128,133,135,138,145,147,154,156,157,166,167,169,171,172,174–176,184,185,188,210]
Leukopenia	40/86 (46.5%)		[6,8,14,18,39,40,52,64,128,133,156,157,176,182,185,186]
Leukocytosis	16/86 (18.6%)	A normal white count was observed in 30/86 (34.9%) patients	[9,10,22,35,57,110,113,117,150,154,171–175,186]
Neutropenia	13/86 (15.1%)		[23,39,64,76,86,104,128,136,140,157,186,188,189]
Thrombocytopenia	20%	Platelet count was not documented in the majority of cases	[1]
Thrombocytosis		Platelet count was not documented in the majority of cases	[60]
Absence of eosinophils	17/86 (19.8%)		[45,51,78,83,112,127,145,152,186,188]
Lymphocytopenia (defined as <1000 lymphocytes/mm3)	27/77 (35.1%)	Normal lymphocyte count was found in 50 (64.9%) patients	[5,7–10,14,15,31,38,39,41,42,44,46,54,57,58,64,76,78,83,93,104,112,113,115–119,128,135,142,145,150,155,159,167,171–175,179,182–184]
Low (<3%) or absent peripheral B cells	66/75 (87%)	Normal B cells in 9/75 patients (13%)	[1,6–9,14,15,18,21,26,27,29–31,36,38–40,42,44,46,51,52,54,56,58,60,61,73,74,77,78,83,94,98,103,105,112,114,116,119,124,127,128,138,211]
Low T cells	6/40 (15%)	28 patients (70.0%) had normal number of T-cells. 6 (15%) patients had elevated T cells	[7–9,14,21,23,27,29–32,38,44–46,46,52,56,59,87,99,103,105,112,114,115,115,116,127,128,134,138]
Low CD4 count (<360/ul or <36%)	41/56 (73.2%)	15/56 (26.8%) had normal CD4 count	[1,2,8,14,15,18–20,26,27,30,36–38,40,42,51,54,64,65,69,74,75,78,82,84,86,98,99,104,112,113,115,116,118,127,133,211] [1,2,7,9,15,29,44,52,56,58,73,94,99,101,115]
A high CD8 count (>1000 cells/μl or >35%)	22/40 (55%)	Normal CD8 count in 14/40 patients (35%) and a low CD8 count in 4/40 patients (10%)	[1,2,7–9,14,27,29,30,37,38,40,44,52,54,56,58,65,69,73–75,78,82,84,94,98,99,101,104,112,113,115,115,122,127,133,134]
Low CD4/CD8 ratio	51/67 (76.1%)	An etiology for the low CD4/CD8 ratio was not	[15,18,27,39,40,44,54,60,64,78,84,94,124]

Laboratory data	Number of patients with available data (%)	Comments	References
		determined in 8 cases and in 22 (51.2%,22/43) patients was caused by a low CD4 count, in 11 patients (25.6%, 11/43) by a combination of low number of CD4 cells and high number of CD8 cells, in 5 (11.6%, 5/43) patients was caused by an increased number of CD8 cells and in 5 patients the CD4 and CD8 cell counts were normal but their ratio was low(11.6%, 5/43).	
Low NK cells	4/7 (57.1%)	NK cells were elevated in 3 other patients.	[2,9,30,42,77,78,84]
Hypogammaglobulinemia	152/152 (100%)	Details on the deficiency of subtypes of globulins were documented in 110 cases but not in 42 other cases. Panhypogammaglobulinemia with absence of all globulins (IgG, IgA, and IgM) was found in 82/110 patients (74.5%). Low levels of only IgG was found in 10/110 patients (9.1%) and low levels of only IgA in 2/110 patients (1.8%). 8/110 patients (7.3%) had both low IgA and IgG and 5/110 patients (4.5%) had low IgG and IgM and 3/110 patients (2.7%) low IgA and IgM. 3 patients had documented absent IgD globulins and 9 patients had documented absent IgE levels. Immunoglobulins subgroups were specified in only one case with a normal IgG1 but depressed levels of IgG2, IgG3 and IgG4.	[2,5–10,13–15,18,26,27,30,35,39–41,42,44,46,51,52,54,57–60,64,65,73,77,78,81–83,85,87,93–95,98,102,110,112,113,115–117,119,124,127,128,133–136,138,141,142,145,146,154,155,157,167,171,174,175,182,184,211]
Normal complement levels	10/12(83.3%)	Levels were high in one case[133] and low in one case.[13]	[37,58,78,83,116,119,127,128,148,189]

Laboratory data	Number of patients with available data (%)	Comments	References
Autoimmune antibodies	20/36 (55.6%)	The most common autoimmune antibodies were antinuclear antibodies which were present in 11 cases (55%), followed by anti-striated antibodies in 6 cases (30%) and anti-thymic antibodies in 4 cases (20%). Other autoantibodies reported included anti-DS-DNA antibodies, anti-acetylcholine receptor antibody anti-ENA, anti-RNP, p-ANCA, antimicrosome, anti-thyroglobulin, anti-intrinsic and anti-parietal antibodies.	[1,13,26,54,76,103,112,114,115,128,133,145,155,166,168,173,175,180]
Abnormal skin tests	Absent skin reactivity to both PPD and at least 1 control antigen was noted in 28/43 patients (65.1%)	11 patients (25.6%) had normal control reactivity, [15,39,145] 2 patients (4.7%) had a positive PPD skin test with negative controls,[127,147] and 2 patients (4.7%) had positive skin tests for at least one control antigen but negative test for PPD or other controls [154,182].	[26,42,44,45,57,58,64,74,77,83,93,98,105,112,113,118,119,128,133,138,146,155,157,166–168,172]

Abbreviations: PPD: purified protein derivative.