AN ESSAY
ON
JUVENILE RHEUMATOID ARTHRITIS
(STILL'S DISEASE)

A THESIS
SUBMITTED FOR PARTIAL FULFILLMENT OF MASTER
DEGREE IN PEDIATRICS

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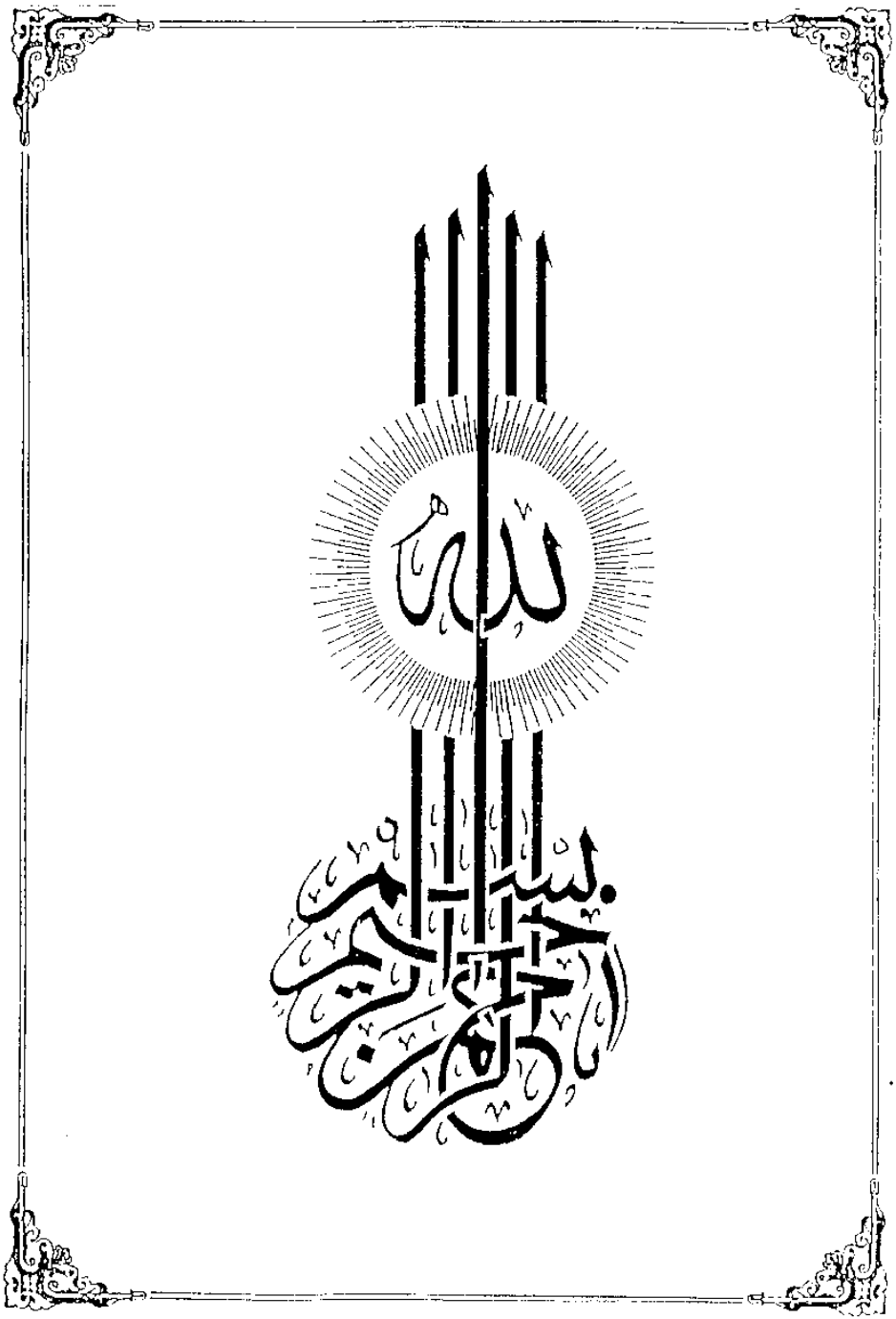

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INTRODUCTION

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Juvenile rheumatoid arthritis (JRA)is afrequent pediatric connective tissue disease. It is one of the more common chronic illnesses of childhood and a leading cause of disability and blindness (Cassidy, 1981).

JRA is the name in the United States of America and its synonyme in the United Kingdom is Juvennile Chronic Arthritis (JCA). Still's disease often referrs to the acute systemic onset of the disease although this emponym is also used for JRA in general (Schaller, 1981).

JRA is a systemic rheumatic disease with protean clinical manifestation that begin before puberty. It affects large joints as the knees, wrists and ankles more than small ones. General affection as pericarditis, hepatitis and uveitis are commoner in JRA than in adult onset R.A. Subcutaneous nodules, rheumatoid seropositivity are unusual (Cassidy, 1981).

Incidence:

It is not a rare disease. Very few published data have centered sepcifically on its prevalence. No race or climate is excluded from its attack (Baum, 1977).

In the United States it is estimated that there are one quarter of a million children with JRA (Calabro and Marchesano, 1967).

In Michigan the data provided a minimal estimate of the incidence of disease: 0.01 percent of children at risk per year (Sullivan, et al., 1975).

In Finland, Leaaksonen reported a similar figure of approximately 0.008 per cent of children under the age of 15 affected per year (Laaksonen, 1966).

History:

In 1897 Still presented a masterly description of both the acute and chronic forms of rheumatoid arthritis in children. Still gave detailed description of 19 children with chronic arthritis whom he had seen during his training in pediatrics and pathology at the Hospital for Sick Children, Great Ormond Street, London. He noted a diversity of disease manifestations in these children: 12 patients had disease characterized by arthritis and extra-articular manifestations as splenomegaly, lymphadenopathy, fever, and anaemia. 6 patients had disease which resembled adult rheumatoid arthritis, and 1 patient had chronic rheumatism. Based on these observations he suggested that chronic childhood arthritis represented more than one disease.

In the ensuing 80 years there has been considerable confusion and controversy about the matter of chronic childhood arthritis, its definition and its relationship to other forms of arthritis (Schaller, 1981).

AIM OF THE WORK

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The aim of this work is to write an essay about (Juvenile Rheumatoid Arthritis). Our review will include.

* Etiology.
* Diagnosis.
  - Clinical picture with reference to different subtypes.
  - Investigations.
* Differential diagnosis.
* Treatment.

ETIOLOGY

The etiology of rheumatoid arthritis and the mechanism for perpetuation of chronic synovial inflammation in the disease are unknown. two frequent hypotheses are that the disease results from:

An infection with as yet unidentified micro-organisms

OR

Autoimmunity to unkown stimuli (Schaller, 1983)

AUTOIMMUNITY

An enormous amount of evidence supports the concept that the pathology of rheumatoid arthritis has its basis in inflammatory response involving the immune system. There are indications that rheumatoid arthritis is an autoimmune disease (Bennet, 1981):

A) The Immunological Hyperactivity of the Rheumatoid Synovium:

There is marked infiltration of the synovium with immune cells. The synovium is packed with lymphocytes and plasma cells. The rheumatoid synovial fluids contain large numbers of blood cells, predominantly neutrophils. The magnitude of this cellular response is not sufficiently appreciated until one considers that a knee joint with a 10 c.c effusion actively contains 100 million neutrophils having 3-4 hours half-life, thus 50 million neutrophils would be entering the articular cavity six times daily to participate in the inflammatory reaction (Zvaifler, 1976).

In vitro experiments employing radiolabled amino acids indicate that immunoglobulin synthesizing capacity of rheumatoid synovial membrane is similar to that of lymph nodes or splenic tissue(Smiley,et al.,1968).

B) Reduced complement activity:

The complement components wheh studied in synovial fluid of actively inflamed rheumatoid joints were found decreased.The decrease could result from one of several mechanisms:-

- Selective destruction of a single component.
- An increase of an inhibitor of the complement.
- Consumption of the late components of complement through an alternate pathway.

The overall pattern of complement depletion has been determined.C1,C4,C2 and C3 are all depressed and depressed proportionately.Rheumatoid fluids also show evidence of the presence of C3 convertase and the trimolecular complex $C\overline{567}$.These results are most consistent with immune activation of complement sequence (Zvaifler,1976).

C) Presence of immune complexes:

Evidence for the presence of immune complexes in synovial fluid is very strong.A factor resembling soluble antigen-antibody complexes has been reported in three quarters of rheumatoid synovial fluids examined by the measurement of histamine release from guinea pig lungs (Baumal and Broder,1968).

Experimental synovitis has been produced by injection of autologous intact IgG or the F_c or F_{ab} fractions into the uninvolved knee joints of patients with seropositive arthritis (Hollander,et al.,1968).