



Comparative Study of Different Regimens in Treatment of Immune Thrombocytopenic Purpura in Children

Thesis

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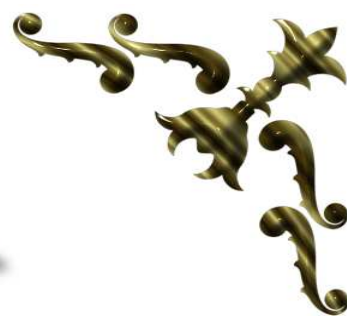
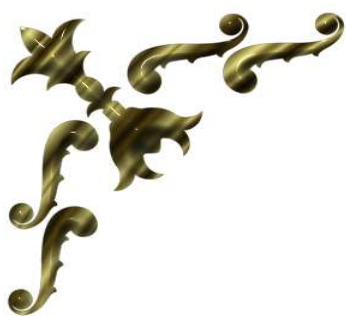
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وَقُلْ اَعْمَلُوا فَسَيَرَى اللّٰهُ
عَمَلَكُمْ وَرَسُولُهُ وَالْمُؤْمِنُونَ



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List of Abbreviations

| | |
|----------------|--|
| • 111-I | Indium-111 |
| • AA | Arachidonic acid |
| • ACTH | Adrenocorticotrophic hormone |
| • ADP | Adenosine diphosphate |
| • ANA | Antinuclear antibody |
| • ATP | Adenosine triphosphate |
| • BCSH | British Committee for Standards in Hematology |
| • B-TG | B-thromboglobulin |
| • C3b | Complement 3b |
| • Ca | Calcium |
| • CD | Cluster of differentiation |
| • cITP - csITP | Chronic ITP, chronic severe ITP |
| • CNS | Central nervous system |
| • CR | Complete response |
| • CSF | Cerebrospinal fluid |
| • CVI | Common variable immunodeficiency |
| • EDTA | Ethylenediaminetetraacetic acid |
| • Epi | Epinephrine |
| • GM-CSF | Granulocyte-macrophage-colony stimulating factor |
| • GP | Glycoprotein |
| • HDMP | High dose methyl prednisolone |
| • HELLP | Hemolysis, elevated liver enzymes, low platelets |
| • HIV | Human immunodeficiency virus |
| • HUS | Hemolytic uremic syndrome |
| • ICH | Intracranial hemorrhage |
| • IG | Immunoglobulin |
| • IL | Interleukin |
| • INF | Interferon |
| • ITAM | Immunoreceptor tyrosin-based-activation motifs |
| • ITIM | Immunoreceptor tyrosin-based-inhibitory motifs |
| • ITP | Immune thrombocytopenic purpura |

| | |
|--------|-------------------------------------|
| • IVIG | Intravenous immunoglobulin |
| • LAT | Lymphotoxin |
| • LDH | Lactate dehydrogenase |
| • MHA | Microangiopathic hemolytic anemia |
| • MMR | Measles, mumps, rubella |
| • NR | No response |
| • PAF | Platelet activation factor |
| • PDGF | Platelet-driven growth factor |
| • PF | platelet factor |
| • PG | Prostaglandine |
| • PL1A | Platelet antigen 1A |
| • PLT | Platelet |
| • PR | Partial response |
| • PRP | Platelet rich plasma |
| • PTP | Post transfusion purpura |
| • RES | Reticuloendothelial system |
| • SLE | Systemic lupus erythrematosis |
| • TG-b | Transforming growth factor b |
| • TH | T-helper cell |
| • TMA | Thrombotic microangiopathies |
| • TNF | Tumor necrotic factor |
| • TPO | Thrombopoietin |
| • TR | T –regulatory cell |
| • TTP | Thrombotic thrombocytopenic purpura |
| • TXA2 | Thromboxane A2 |
| • vW | von Willebrand's factor |

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Introduction



Introduction

Immune thrombocytopenic purpura (ITP) in children is usually a self limiting disorder presenting most commonly with a short history of purpura and bruising in children of either sex between the ages of 2 and 10 years of age. The incidence is at about 4 per 100 000 children per year. It may follow a viral infection or immunization and is caused by an inappropriate response of the immune system. Autoantibodies against platelet surface glycoproteins (particularly IIb/IIIa) can commonly be detected (60-70%), but are of no prognostic significance and this is not a useful diagnostic test (**Maggs et al, 2001**).

A wide range of therapeutic regimens are currently in use, including observation alone, as the majority of children recover within 4-6 months regardless of treatment. A growing understanding of the pathophysiology of acute ITP in children has not solved the controversy of treatment, but has clarified the mechanism of action of the most frequently used agents in chronic ITP (**Nugent et al, 2006**). Currently, there is no single optimal management for the child who is newly diagnosed with ITP. Most hematologists in the United States choose to treat a child with a platelet count $<10,000 \times 10^3/\mu\text{L}$ or with mucous membrane bleeding. However, observation and education are appropriate for the child with mild thrombocytopenia and no clinical bleeding. Initial treatment options for childhood ITP include IVIG, anti-Rh immunoglobulin ("anti-D"), steroids (oral or IV), or combination therapy (**Beardsley et al, 2006**).

About 20% of the children diagnosed with acute ITP will run a chronic course. Only in a minority of these, platelet-count-enhancing