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Outcome of children with idiopathic thrombocytopenic purpura, based on original material

Idiopathic thrombocytopenia purpura (ITP) is a common pediatric disease with various phenotypes. An inappropriate immune response induced by virus infection and other antigens, including autoimmunity, may result in platelet-associated antibodies. As a consequence of the interaction the antibody coated platelets are destructed in the spleen, which may cause thrombocytopenia (platelet count < 150 x 10^9/L). ITP is subdivided into an acute and chronic form based solely on the duration of thrombocytopenia (3, 5, 6, 9).

The typical presentation of acute ITP is an abrupt onset of bruises and petechia in an otherwise healthy child. An antecedent viral illness is often reported in the weeks prior to diagnosis. Less frequent presentation includes epistaxis. In a small number of patients acute ITP accompanies infection with rubella, mumps, varicella, Epstain-Barr virus, cytomegalovirus, HIV. The incidence of life-threatening hemorrhage in acute ITP is very rare (less than 1%). All reported life-threatening bleeding, which has occurred in patients with platelet counts < 20 x 10^9/L (1, 5).

Chronic ITP occurs in 10–20% of children with acute ITP and is defined as thrombocytopenia that persists for more than 6 months. Chronic ITP is more common in older children (>13 years of age). The initial clinical and laboratory features cannot distinguish between acute and chronic ITP and do not predict which patient will develop chronic ITP (8).

There are the two major therapeutic options for treatment of acute ITP such as steroids and intravenous immunoglobulin. For children with chronic ITP the treatment options are splenectomy or immunosuppressive therapy. Some physicians would recommend watchful waiting because with time most children with chronic ITP show spontaneous improvement in bleeding manifestation and usually platelet count as well (3, 4).

The aim of the study was to analyse the course of disease and outcome of children with acute and chronic ITP.

MATERIAL AND METHODS

The study comprised 52 patients with ITP, who were treated in the Department of Pediatric Hematology and Oncology in Lublin from 1998 to 2002. In the study the course of disease, therapeutic methods and results of the treatment were evaluated.

RESULTS

Of the 52 patients with ITP there were 28 girls and 24 boys aged from 12 months to 17 years (mean 9.8 years). Viral infection was reported in ten patients (rubella in 4, mumps in 3, varicella in 3). Megacaryocyte thrombocytopenia was diagnosed in 50 patients and amegacryocyte in two of them. Methods of therapy pictures Table 1. Spontaneous recovery was observed only in three patients (5,7%). Good response to treatment was found in 40 (76.9%) patients. The follow-up is from three months to three years (mean 19.8 months). Chronic ITP occurred in nine (17.3%) patients. Out of nine patients, six had a splenectomy, which was performed after 12 months' conventional treatment with good response. Three children of this group are observed only in the outpatient clinic (their parents did not agree to surgery). In these patients the level of platelet count varies from 35 x10^9/L to 70 x 10^9/L.

Table 1. Therapeutic methods, which were applied in children

Therapeutic methods	N	%
Prednison 2 mg/b.w.	33	63.4
Immunoglobulin 400 mg/b.w.	1	1.9
Prednison + immunoglobulin	16	30.7

DISCUSSION

Idiopathic thrombocytopenic purpura in children is usually a benign and self-limited disease (9). Two concerns dominate the initial evaluation and treatment schedule: 1) Does the child with the symptoms of ITP have acute leukemia, aplastic anemia or other serious diseases instead? 2) Is the child at immediate risk of life-threatening hemorrhage? (3, 6). Both concerns are fueled by the marked reduction platelet count that accompanies the physical signs of bleeding. The clinical diagnosis of ITP includes medical history, physical examination, complete blood count and peripheral blood smear and bone marrow puncture to exclude other causes of thrombocytopenia (2). After diagnosis we decided about therapeutic option. Corticosteroids are one of the two major treatment options. Steroids increase vascular stability and platelet production, decrease antiplatelet antibody production (3). In our study, 33/52 patients who were treated prednison achieved good response. In 16/52 patients response to steroid therapy was poor and immunoglobulin was applied additionally. Out of 16 patients, seven had good response to combined treatment and nine had diagnosed chronic ITP. Some physicians fear splenectomy as the treatment of choice in these patients and we agree with this opinion. All our children with chronic ITP having splenectomy respond fully. Post-splenectomy septicemia is prevented by prophylactic penicillin, vaccination against encapsulated organism and most especially, medical attention and administration antibiotics at times of illnesses (1, 9). Out of nine patients with chronic ITP, three had not splenectomy (lack of parents' agreement) and these children require more often control of the level of platelet count.

Despite generally moving in the same direction, the management of ITP does differ markedly worldwide and there is clear need for cooperative studies to learn more about diagnosis, management and prognosis (2).

CONCLUSIONS

1. The benefits of splenectomy in markedly symptomatic chronic ITP far outweigh the risks.

2. The majority of patients with acute ITP had good response after corticosteroids therapy.

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SUMMARY

Idiopathic or immune thrombocytopenia purpura (ITP) is a common hematologic disease in children. ITP is subdivided in an acute and chronic form based solely on the duration of thrombocytopenia. The aim of the study was to analyse the course of disease and outcome of children with acute and chronic ITP. The study enrolled 52 patients with ITP, who were treated in the Department of Pediatric Hematology and Oncology in Lublin from 1998 to 2002. Of the 52 patients with ITP there were 28 girls and 24 boys aged from 12 months to 17 years. Out of the 52 patients, 3 patients had spontaneous recovery, 33 were treated with steroids, one with immunoglobulin, others with combined therapy. Good response to treatment was found in 40 patients. Nine children developed chronic ITP. Out of 9 patients, 6 had splenectomy with good result and 3 are only observed. The benefits of splenectomy in markedly symptomatic chronic ITP far outweigh the risks. The majority of patients with acute ITP had good response after corticosteroids therapy.

Wyniki leczenia dzieci z idiopatyczną małopłytkowością samoistną w materiale własnym

Samoistna idiopatyczna małopłytkowość (ITP) jest jedną z najczęściej występujących chorób hematologicznych u dzieci. ITP może występować w postaci ostrej i przewlekłej. Celem pracy była analiza przebiegu klinicznego i wyników leczenia dzieci z ostrą i przewlekłą idiopatyczną małopłytkowością samoistną. Badaniem objęto 52 pacjentów z ITP leczonych w Klinice Hematologii i Onkologii Dziecięcej AM w Lublinie w latach 1998–2002. W badanej grupie było 28 dziewczynek i 24 chłopców w wieku 12 miesięcy–17 lat. W leczeniu zastosowano sterydy u 33 pacjentów, immunoglobulinę u jednego i terapię skojarzoną u 16. Dobrą odpowiedź na leczenie uzyskano u 40 dzieci. Dziewięciu pacjentów rozwinęło przewlekłą ITP. Z tej grupy sześcioro dzieci miało wykonaną splenektomię, a trójka pozostaje jedynie w obserwacji. Można wnioskować, że korzyści ze splenektomii u dzieci z przewlekłą IPT przeważają nad ryzykiem. Większość pacjentów z ostrą ITP dobrze zareagowała na sterydy.