

## Childhood Idiopathic Thrombocytopenic Purpura: A Retrospective Analysis Of Clinical Features And Response To Treatment

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### Summary:

**Background :** A retrospective study was done in the Pediatric ward /AL-Kadhimiya Teaching Hospital on cases diagnosed as idiopathic thrombocytopenic Purpura admitted since the first of January 1992 – the end of December 2004.

**Aim of the study :** The objective of this paper is to review the natural history of idiopathic thrombocytopenic Purpura , presenting feature and response to treatment.

**Patients and methods :** The review included age ,sex , clinical presentation , physical finding , complications , investigation ,treatment and course of the diseases .

**Results:** total number of the patients were 65, peak age was between (2 -5) years, 40 cases (61.53 %). Females were affected more than Males with male: female ratio of 1: 2.09 .There was no seasonal variation and all of the cases were preceded by viral upper respiratory tract infection .The main presentation was petechiae and ecchymosis, were found in all of them (100%) followed by epistaxis, 43 cases (66.15 %). In the majority the platelet count was  $< 20.000 \times 10^9 /L$ , 32 cases (49.23 %) and the hemoglobin level was between 10 – 11gm /dl in most of them, 43 cases (66.15%). Steroid was the first line treatment, response occur within 2 – 3 weeks in the majority, 51 cases (78.46%). Chronic idiopathic thrombocytopenic Purpura occurred in 3 cases only (4.61 %). Splenectomy was done in 2 of them (66.66%). No mortality detected.

**Conclusion :** Idiopathic thrombocytopenic purpura is a mild disease with complete recovery in the majority of patients

**Key words:** Thrombocytopenia, Purpura, Children

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### Introduction:

Immune thrombocytopenic Purpura (ITP) due to production of anti – platelet antibodies, is a clinical syndrome in which decreased number of circulating platelets manifest as bleeding tendency <sup>(1)</sup>. Typically benign, acute self limiting illness usually occurring after an infectious diseases <sup>(2)</sup>. Despite very low platelet count is rarely complicated by serious bleeding <sup>(3)</sup>. Mortality is rare mostly due to intracranial hemorrhage which occurs in about 0.1% of children with a platelet count  $< 20.000 \times 10^9 /L$  <sup>(4)</sup>.

### Patients And Methods:

A retrospective study was made for children (1 – 16 years) admitted to the Pediatric ward / AL – Kadhimiya Teaching Hospital from the first of January 1992 - 31 of December 2004 with idiopathic thrombocytopenic Purpura . The review include 1-age , 2-sex , 3-preceded upper respiratory tract infection ,4- preceded vaccination , 5-season of presentation , 6-clinical presentation , 7-physical finding ,8- complication , 9-hemoglobin level and platelets count ,10- bone marrow finding , 11-treatment given , 12-response to therapy , 13-course of the disease whether acute or chronic.

Complete remission is defined as an increase in the platelet count  $> 150.000 \times 10^9 /L$  <sup>(5)</sup>. Acute

idiopathic thrombocytopenic Purpura are cases with typical presentation of bleeding episodes lasting for few days or weeks but no longer than 6 months <sup>(6)</sup> supported by low platelet count in the peripheral blood film and bone marrow finding of normal or increased megakaryocytes count<sup>(6)</sup>. Chronic idiopathic thrombocytopenic Purpura is defined as platelet count of  $< 150.000 \times 10^9 /L$  persisting for more than 6 months from the onset of illness <sup>(7)</sup>.

### Result:

The study was based on 65 cases . Age range was ( 1 – 16 years) with a peak forty patients ( 61.53%) in the age range ( 2 -5 years) as shown in ( table – 1).

Females were affected more than males , 44 cases ( 67.69%) , 21 cases ( 32.30%) respectively with male : female ratio equal to 1: 2.09 .

There was no seasonal variation in relation to the onset of the disease , cases were reported through out the year. All of them gave a history of preceded upper respiratory tract infection ( 100%) 3 – 4 weeks prior to the onset of the disease . There was no preceding history of vaccination .

The main clinical finding were skin manifestation in the form of petechiae , purpura and ecchymosis seen in all of the patients (100%) , followed by epistaxis ,recorded in 43 cases ( 66.15%) , as shown in ( table -2 ) .

Thirty two patients ( 49.23%) , had platelet count  $< 20.000 \times 10^9 /L$  , as shown in ( table – 3

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).Hemoglobin level was in the range of (10 -11g/dl) in 43 patients( 66.15%) as shown in (table – 4). Bone marrow was performed in all of the cases , reveal either normal or increased megakaryocytes count , no evidence of abnormal cells.

Oral prednisolone was the main line of treatment , it was given to all of the patients . Complete response within 2 -3 weeks of treatment was found in 51 cases ( 78.46 %) whereby the platelet count increase to  $\geq 150.000 \times 10^9/L$  . Partial response with a platelet count between  $120.000 - 150.000 \times 10^9/L$  was found in 10 cases ( 15.39%) . one case ( 1.54%) was not responding to the usual dose of steroid ( 2 mg / kg / day ) with massive gastrointestinal bleeding in the form of clots per rectum which continued even after increasing the dose to ( 5 mg/kg/day ) .Splenectomy was done in the acute phase of the disease as an emergency and obligatory treatment to save the life of this patient. Three patients (4.61%) were not responding to treatment with a persistently low platelet count  $< 20.000 \times 10^9/L$  for 6m – 1 year recorded ( chronic idiopathic thrombocytopenic Purpura ) . High dose dexamethasone  $40\text{mg}/\text{m}^2$  / day for 4 days every 28 days lasting for 6 months was given to two of them . Response was good with persistence of platelet count  $>150.000 \times 10^9/L$  lasting for 6 months after treatment seen in both but eventually the platelet count decline again and finally Splenectomy was done for both of them with complete remission .Immunoglobulin was not used because it was not available in the hospital . No death was recorded.

**Table – 1 –Age Distribution of 65 patients with idiopathic thrombocytopenic purpura**

Age / year	No.	%
$\leq 1$	1	1.54
2 – 5	40	61.53
6 – 9	12	18.47
10 – 13	7	10.77
$> 13$	5	7.69

**Table – 2 –The Clinical Features of 65 patients with idiopathic thrombocytopenic purpura**

Clinical features	No.	%
Petechiae	65	100
Ecchymosis	65	100
Epistaxis	43	61.15
Gum bleeding	28	43.07
GIT bleeding / rectum	3	4.61
Hematuria	2	3.07
Intracranial hemorrhage	0	0
Pallor	8	12.30
Lymphadenopathy	0	0
Hepatomegaly	0	0
Splenomegaly	2	3.07

**Table – 3 –Platelet Count of 65 patients with idiopathic thrombocytopenic purpura**

Platelet Count $\times 10^9/L$	No.	%
$< 20.000$	32	49.23
$20.000 - 50.000$	21	32.30
$> 50.000$	12	18.47

**Table – 4 –The Hemoglobin Level of 65 patients with idiopathic thrombocytopenic purpura**

Hemoglobin level g/dl	No.	%
$< 10$	2	3.07
10 – 11	43	66.15
$> 11$	20	30.78

#### Discussion:

In this study forty patients were in the range of ( 2 – 5 years ) of age which is in agreement of Sandler SG et al <sup>(1)</sup>, AL – Nadawi MN et al <sup>(8)</sup> and Nugent et al <sup>(9)</sup> studies .The disease affecting females more than males which is also in agreement of AL – Nadawi MN et al in Iraq <sup>(8)</sup> and Mantakis E et al study in USA <sup>(10)</sup>.

There was no seasonal variation , in comparison with a study done in Argentina , greatest frequency was found in Spring ( 36.3 % ) , (Donato H et al) <sup>(11)</sup> .In 100 patients preceded viral upper respiratory tract infection were recorded with no history of preceded vaccination .In Argentina preceding viral respiratory tract infection was seen in (68.6%), (Donato H et al) <sup>(11)</sup> while prior vaccination was reported in 1/3 of the patients in a study done in Lebanon , (Moussalem M. et al) <sup>(12)</sup>.

Petechiae and ecchymosis were recorded in all patients followed by epistaxis ( 66.15%) which is similar to AL – Nadawi MN et al study , ( 100% , 48.4% ) respectively <sup>(8)</sup> . In Argentina , skin manifestation reported in ( 99%) and epistaxis in (39.2%)( Donato H et a) <sup>(11)</sup> . In Lebanon epistaxis seen in (10%) (Moussalem M. et al) <sup>(12)</sup> while in Texas major bleeding like epistaxis reported in ( 17%) ( Medeiros D et al ) <sup>(13)</sup> , and in ( 2.5 %) in Germany ( Sutor AH et al ) <sup>(14)</sup> .Gum bleeding occur in (28%) of cases, a finding similar to AL – Nadawi MN et al study in Iraq (30.9%)<sup>(8)</sup>. Massive gastro- intestinal bleeding found in ( 4.1%), in a study done in United Kingdome gastro- intestinal bleeding was found in ( 5%) of children( Bolten – Maggs P ) <sup>(3)</sup> . Hematuria reported in ( 3.07%) a finding less than what was reported before ( 8.2%) ( AL-Nadawi MN et al ) <sup>(8)</sup> . Intracranial hemorrhage

was not encountered in this study . In Japan it was reported in ( 0.52%)( Iyori H et al )<sup>(15)</sup> and usually occur within three days of illness<sup>(16)</sup> .

Pallor had been noticed in ( 12.30%) mainly in those with massive gastro- intestinal bleeding and epistaxis , anemia is unlikely to be seen in idiopathic thrombocytopenic Purpura unless there is massive bleeding<sup>(1)</sup> . The majority had a platelet count  $<20.000 \times 10^9/L$  ( 49.23%) which is in agreement of Erduran E et al study<sup>(4)</sup> and Rosthoj S et al study<sup>(17)</sup> .

Although routine performance of bone marrow examination for the diagnosis of idiopathic thrombocytopenic Purpura is not necessary provided that a thorough history and physical examination are performed and that complete blood count , peripheral blood smear show no abnormalities apart from thrombocytopenia , still the value of bone marrow investigation in idiopathic thrombocytopenic purpura remains unresolved<sup>(18)</sup> .Most physician continue to recommend this investigation before corticosteroid are administered<sup>(2)</sup> .

Steroid ( oral prednisolone ) was the main mode and first line of treatment given to all of the patients , complete remission was observed in ( 78.46%) , a finding similar to Erduran E et al<sup>(4)</sup> and Andres E et al studies<sup>(19)</sup> .

Chronic idiopathic thrombocytopenic Purpura was reported in ( 4.61%) , a finding less than other studies being ( 10%) in Lebanon ( Moussalem M et al )<sup>(12)</sup> , (25%) in Canada<sup>(20)</sup> ( Ahmed S et al )and in (47%) of children aged 120 months and above( Sandoval C et al )<sup>(21)</sup> .High dose dexamethasone was applied as a second line of treatment in those patients with complete remission lasting for 6 months, a finding higher than Pamuk G et al study in Turkey whereby (44.4%) of patients showed complete remission when high dose steroid was used as a second line treatment<sup>(22)</sup> . Splenectomy was associated with complete remission , in comparison with Pamuk G et al study ,complete remission after splenectomy was obtained in ( 68.4%) in Turkey<sup>(22)</sup> and in ( 63%) of patients in Spain ( Diaz – Conradi A et al )<sup>(23)</sup> .it was found that an initial increase in platelet count after steroid bolus is a good indicator for favorable response to splenectomy<sup>(24)</sup> .Younger age and peak postsplenectomy platelet counts were significantly associated with a favorable response to splenectomy<sup>(25)</sup> .It is concluded from this study that idiopathic thrombocytopenic Purpura is not a serious disease in the majority with a good prognosis.

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