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ณ ห้องประชุมพจมาน-ทักษิณ ชินวัตร ตึกพัชรกิติยาภา
โรงพยาบาลพระมงกุฎเกล้า

Anemia in Pregnancy : Trends and Etiology in Thailand

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Anemia in pregnancy is an important health problem in the world especially in developing countries. It remains a major contributing factors to maternal morbidity and mortality and is associated with high perinatal mortality rate. In Thailand the prevalence of anemia during pregnancy reported from few university and provincial hospitals varied between 50% to 21% in the year 1970 to 1983. Ministry of Public Health reported the prevalence of anemia in pregnancy in Thailand as high as 28.0% to 17.8% in 1987 and 1991 accordingly. Those reports were from the hospital based data with different methods and standard of laboratory testing for anemia. Moreover there has not been any study to identify the etiology of anemia in Thailand. The objectives of this study is to identify the prevalence of anemia during pregnancy in Thailand and its main common causes. The study design was a cross sectional descriptive study. The target population was the pregnant women attended antenatal care for the first time from six Mother and Child Hospitals in 4 regions throughout Thailand. The standardized laboratory testings consist of hematocrit as a screening test with additional testing of serum ferritin and hemoglobin typing in anemia pregnant women whose hematocrit less than 33%. 624 of 6,536 pregnant women in the study were anemic with the prevalence rate of 9.6%. The highest prevalence at 16% was in the north-east region and the lowest at 4% was in Bangkok and upper-north region. 97% of anemic pregnant women were in mild state of anemia with hematocrit level between 32.9-27.0%. The main causes of anemia were iron deficiency anemia, thalassemia syndromes and Traits, and the combination of the first two causes with the rate of 33%, 25% and 25% accordingly. As a result the iron deficiency anemia and thalassemia syndromes and traits are the two major causes of anemia in Thailand comprised of 5.5% and 4.8% of all pregnant women. Approximately 2/3 of anemia pregnant women in the south, central and lower north region are caused by iron deficiency anemia, whereas 2/3 in the north-east, and Bangkok are caused by thalassemia syndromes and traits. Thalassemia syndromes and traits in Thailand comprise of heterozygote 73% (Hb E, α -1, β -Thalassemia traits) and homozygotes 27% (Hb E disease, β/β , β /E, Hb H disease, Hb Cs, AE Barts and Hb Anantaraj). Hb E disease and traits are the most common type.

Key Words : ● Anemia ● Pregnancy ● Thalassemia ● Iron deficiency

Prevention and Control of Severe Thalassemia Syndrome : Chiang Mai Strategy

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เหตุผลและความเป็นมา : โรคโลหิตจางธาลัสซีเมียได้แก่ Hb Bart's hydrops fetalis (homozygous α -thalassemia1), homozygous β -thalassemia และ β -thalassemia Hb E disease เป็นโรคร้ายแรงทางพันธุกรรม การดูแลรักษาทำให้เกิดการเปลี่ยนแปลงทางเศรษฐกิจต่อครอบครัวประเทศชาติอย่างยิ่ง การให้บริการ genetic counseling ที่เหมาะสมกับหญิงฝากครรภ์น่าจะมีประโยชน์ในด้าน health economics

วัตถุประสงค์ : เพื่อศึกษาถึงแนวทางและกลวิธีที่เหมาะสมในการควบคุมและป้องกันโรคโลหิตจางธาลัสซีเมียชนิดร้ายแรงแบบครอบครัว

ตัวอย่าง : เป็นหญิงที่มาฝากครรภ์ ณ โรงพยาบาลมหาสารคามเชียงใหม่ ระหว่างเดือนมกราคม 2539-เมษายน 2540 จำนวน 4,103 ราย ซึ่งมีอายุครรภ์ระหว่าง 16-20 สัปดาห์

Chiang Mai Strategy : 1. ตรวจกรองพาหะของโรคธาลัสซีเมียหญิงฝากครรภ์ด้วยวิธี 2 min OF (2 minute osmotic fragility test) และ Hb E ด้วยวิธี Hb E screening 2. ตรวจหาชนิดของพาหะได้แก่ β -thalassemia trait, Hb E trait, homozygous Hb E ด้วยการวัดปริมาณ Hb A₂ และใช้วิธี PCR (polymerase chain reaction) สำหรับ α -thalassemia 1 trait (SEA type) 3. เฉพาะคู่สมรสที่มีความเสี่ยงมีบุตรเป็นโรค homozygous β -thalassemia, β -thalassemia Hb E disease และ Hb Bart's hydrops fetalis จะได้รับการตรวจเลือดจากสายสะดือโดยวิธี cordocentesis เมื่ออายุครรภ์ 18-20 สัปดาห์ 4. ให้การวินิจฉัยโรคธาลัสซีเมียจากเลือดของทารกด้วยวิธี HPLC (high performance liquid chromatography) Hb analysis 5. หากพบว่าทารกเป็นโรคธาลัสซีเมียจะได้รับคำแนะนำและให้การรักษาทางพันธุกรรมที่เหมาะสมต่อไป

ผลการศึกษา : หญิงฝากครรภ์จำนวน 4,103 รายพบมีความผิดปกติจากการตรวจ 2 min OF 1,850 ราย (28.8%) แยกเป็น β -thalassemia trait 5.6%, thalassemia 1 α -trait 7% พบ Hb E 9.6% คิดเป็นความถี่ของยีนพาหะแต่ละชนิดได้ดังนี้ β -thalassemia gene 0.028, α -thalassemia gene 0.035 และ Hb E gene 0.048 ตามลำดับ การเฝ้าระวังในระยะเวลา 1 ปี (พค. 39-เมย. 40) มีหญิงที่ฝากครรภ์จำนวน 3,383 ราย สามารถคำนวณโอกาสการเกิดโรคตามกฎของ Hardy และ Weinberg จากความถี่ของยีนพาหะแต่ละชนิดจะได้คู่สมรสที่มีโอกาสเสี่ยงเกิดบุตรเป็น Hb Bart's hydrops fetalis 16.5 คู่, homozygous β -thalassemia 10.6 คู่, และ β -thalassemia disease 36 คู่ รวม 63 คู่ ซึ่งจำนวนนี้ควรจะตรวจพบทารกเป็นโรค Hb Bart's hydrops fetalis 4.14 ราย homozygous β -thalassemia 2.65 ราย และ β -thalassemia Hb E disease 9 ราย ในความเป็นจริงมีคู่สมรสที่มารับการตรวจจริงในระยะเวลาดังกล่าว 63 คู่ ซึ่งเป็นคู่สมรสเสี่ยงที่จะเกิดทารกเป็น Hb Bart's hydrops fetalis 15, homozygous β -thalassemia 11 และ β -thalassemia Hb E disease 37 และพบทารกเป็นโรคเท่ากับ 4 ราย, 3 ราย และ 9 รายตามลำดับ ซึ่งใกล้เคียงอย่างมากกับความน่าจะเป็นในเชิงทฤษฎี

สรุป : แนวทางและกลวิธีแบบครอบครัวข้างต้นมีความถูกต้องน่าจะนำมาใช้ในระดับประเทศเพื่อการควบคุมและป้องกันโรคโลหิตจางธาลัสซีเมียชนิดร้ายแรงได้

Key Words : ● Thalassemia ● Osmotic fragility ● Cordocentesis

Partial Splenic Embolization in Thalassemia Children with Hypersplenism : A Preliminary Report

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Purpose : To evaluate the result of treatment by partial embolization of the spleen in β -thalassemia and β -thalassemia/Hb E children with hypersplenism.

Materials and Methods : 25 children with β -thalassemia and β -thalassemia/Hb E, and hypersplenism aged 7-13 years were treated by partial splenic embolization during 1994-1996.

Results : There has significant decrease rate of blood transfusion during 2 years follow up from about 6-12 times/year to 1-2 time/year. The splenic size is also significantly decreased within 6 months. No serious immediate complication except 2 cases which have splenic abscess due to massive embolization and finally splenectomy have to be done. Post operative abdominal pain due to inflammation is a common complication in this treatment. The splenic embolization causes 2 weeks of hospitalization compared to 1 week of the splenectomy but the spleen is intact. Serious infection that commonly occurred in patients splenectomized for thalassemia do not occur during 2 years follow up.

Conclusion : This will be an alternative treatment to splenectomy in β -thalassemia and β -thalassemia/Hb E patients.

Key Words: ● Splenic embolization ● Thalassemia ● Hypersplenism

Hereditary Hemochromatosis : Case Report and Review Literatures

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นิเวศน์ นันทจิต, วรวิ ดันติแพทยางกูร, ธเนศ ชิตาพนารักษ์ และ ศตวรรษ ทองสวัสดิ์
หน่วยโลหิตวิทยา, ภาควิชาอายุรศาสตร์, คณะแพทยศาสตร์, มหาวิทยาลัยเชียงใหม่

Hereditary hemochromatosis เป็นโรคทางพันธุกรรมที่พบได้บ่อยในประเทศทางยุโรป แต่มีรายงานน้อยมากในประเทศแถบเอเชีย ผู้รายงานขอเสนอผู้ป่วย 2 ราย ในครอบครัวเดียวกัน คนแรกเป็นผู้ป่วยชายอายุ 43 ปี เป็นโรคเบาหวาน มาโรงพยาบาลด้วยอาการสำคัญว่า สิวคล้ำมากขึ้น และความรู้สึกทางเพศลดลง ผู้ป่วยปฏิเสธประวัติการรับเลือด การดื่มแอลกอฮอล์ และการใช้ยาใดๆเป็นประจำ ตรวจร่างกายพบลักษณะสีผิวคล้ำทั่วไปโดยเฉพาะที่หลังมือและหลังเท้า ลักษณะบ่งชี้ของเพศชาย เช่น หนวดและขนรักแร้น้อยลง ขนาดอัณฑะเล็กลง การตรวจทางห้องปฏิบัติการพบว่าค่าความอิ่มตัวทรานสเฟอร์ริน (transferrin saturation) มีค่า 91% ผลพยาธิวิทยาของเนื้อเยื่อ ตับมีปริมาณเหล็กสะสมเพิ่มมากขึ้น การประเมินทางโลหิตวิทยาไม่พบความผิดปกติในการสร้างหรือทำลายเม็ดเลือดแดงเพิ่มขึ้น ให้การวินิจฉัยว่าเป็น hereditary hemochromatosis นอกจากนี้ยังตรวจพบว่าผู้ป่วยมี HLA B7 จากการสืบค้นในครอบครัวของผู้ป่วยพบว่าน้องชายของผู้ป่วยมีอาการและอาการแสดงคล้ายกับผู้ป่วย แต่อาการน้อยกว่าผู้ป่วย ได้นำน้องชายผู้ป่วยมาตรวจเลือดหาความอิ่มตัวทรานสเฟอร์ริน (transferrin saturation) มีค่า 94% และการตัดตรวจชิ้นเนื้อจากตับ (liver biopsy) พบมีปริมาณเหล็กสะสมมากในเนื้อตับเช่นเดียวกับผู้ป่วย ผู้ป่วยได้รับการรักษาด้วยการเจาะเลือดออก (blood letting) เมื่อทราบแน่ชัดว่าผู้ป่วยเป็นโรคนี้ ระยะต่อมาได้ให้ (iron-chelating agent (desferrioxamine methanesulfonate) แทน เนื่องจากผู้ป่วยมีภาวะซีดหลังจากเจาะเลือดออกหลายครั้ง ปัจจุบันผู้ป่วยมารับการตรวจติดตามผลที่โรงพยาบาลมหาวิทยาลัยเชียงใหม่ และพบว่าสามารถลดการใช้ยาระดับน้ำตาลในเลือดลงได้

Key Words: ● Hemochromatosis ● HLA B7 ● Blood letting ● Desferrioxamine

Paroxysmal Nocturnal Hemoglobinuria in Chiangmai

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Paroxysmal nocturnal hemoglobinuria (PNH) เป็น acquired clonal disorder ที่มีความผิดปกติเด่นคือ chronic intravascular hemolysis, venous thrombosis และมีความสัมพันธ์กับ aplastic anemia ชัดเจน ซึ่งสามารถอธิบายได้จากการค้นพบ mutation ของ PIG-A gene บน X chromosome ซึ่งทำให้การสร้าง glycosylphosphatidylinositol (GP) anchor ผิดปกติ จึงเกิดการขาด GPI-anchored membrane proteins หลายชนิด โดยเฉพาะอย่างยิ่ง CD 59 และ decay accelerating factor (DAF, CD 55) ที่มีบทบาทสำคัญในการป้องกันการแตกตัวของเม็ดเลือดแดงจากการกระตุ้นระบบคอมพลีเมนต์ รายงานผู้ป่วย PNH ส่วนใหญ่มาจากอเมริกา ฝรั่งเศส จีน สำหรับประเทศไทย ได้มีรายงานเป็นครั้งแรกโดย ศ. นพ.มงคล เครือศรี และคณะ เป็นรายงานผู้ป่วยส่วนใหญ่จากภาคกลางของประเทศ ซึ่งมีลักษณะบางอย่างที่แตกต่างกับรายงานของทวีปอเมริกาและยุโรป ผู้รายงานได้รวบรวมผู้ป่วยจากหน่วยโลหิตวิทยา ภาควิชาอายุรศาสตร์ เชียงใหม่ ซึ่งเป็นศูนย์ในการรักษาผู้ป่วยโรคเลือดเป็นส่วนใหญ่ทางภาคเหนือ พบว่าอายุและเพศที่พบในทั้งสองกลุ่มนั้นไม่แตกต่างกัน พบได้ทุกอายุ กลุ่มที่พบมาก อายุระหว่าง 20-40 ปี อาการแสดงของผู้ป่วยภาคเหนือแสดงลักษณะ hemolytic anemia มากกว่าคือร้อยละ 29.4 ในขณะที่ผู้ป่วยภาคกลางพบเพียงร้อยละ 7.2 อาการเลือดออก (hemorrhagic symptoms) พบในผู้ป่วยภาคกลางมากกว่าคือพบร้อยละ 14.6 และภาคเหนือพบร้อยละ 2.9 และทั้ง 2 กลุ่ม ไม่พบผู้ป่วยที่แสดงอาการของ venous thrombosis ดังรายงานของต่างประเทศเลย

Key Words : ● Paroxysmal nocturnal hemoglobinuria ● PIG-A gene
● Aplastic anemia ● Venous thrombosis

von Willebrand's Disease and Its Response to DDAVP

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Objectives : 1. The descriptive analysis of the clinical manifestations and the evaluation of the diagnostic laboratory testings 2. The comparison between the efficiency of intravenous and intranasal 1-deamino 8D arginine vasopressin (DDAVP) among Thai patients with von Willebrand's disease (vWD).

Study design : The clinical manifestations and the laboratory testings among Thai patients with vWD were analysed. Then, 0.3 µg/kg and/or 150-300 µg of DDAVP were subsequently given. Blood was drawn for the measurement of factor VIII clotting activity (FVIII:C), von Willebrand factor antigen (vWF:Ag) and ristocetin cofactor activity (RiCof) before and at 1,6 and/or 24 hours after the administration of DDAVP. In addition, the bleeding time was checked before and 1 hour after the medication.

Results : Twenty four cases with vWD were included. The two most common bleeding symptoms were ecchymosis (41.7%) and epistaxis (37.5%) which were manifested as early as 2 years old. Although vWD is mostly inherited by autosomal dominance, only 17 cases (70.8%) had a family history of bleeding disorder. The patients could be classified as type I, 22 cases; type II and type III, one each. Consequently, the sensitivity of the laboratory testing for the diagnosis of vWD was as followed : FVIII:C = 43.6%, APTT = 44.7%, BT = 62%, vWF:Ag = 68.1% and RiCof = 89.7% Fifteen patients received intravenous DDAVP, the levels of FVIII:C, vWF:Ag and RiCof reached the maximum increment of 3.63,2.79 and 2.60 times, respectively one hour after the medication. Subsequently, eight out of 15 cases also received intranasal DDAVP, the levels of FVIII:C, vWF:Ag and RiCof reached the maximum increment of 2.74,2.19 and 1.95 times, respectively one hour after the medication. These levels were significantly higher than the pretreatment level ($p < 0.05$) except RiCof via intranasal route. However, the levels of intravenous route were not significantly higher than those of the intranasal route.

Conclusion : The diagnosis of vWD depends primarily upon the manifestation of the bleeding disorder and laboratory testing. The most sensitive test was RiCof. DDAVP either by intravenous or intranasal route effectively increases the levels of FVIII:C, vWF:Ag and RiCof.

Key Words: ● von Willebrand's disease ● DDAVP

การผ่าตัด Sequestrectomy ของกระดูก Ilium ในผู้ป่วย Hemophilia A ที่มี Factor VIII Inhibitor โดยใช้ Recombinant Activated Factor VII ที่โรงพยาบาลจุฬาลงกรณ์

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บทนำ : การผ่าตัดในผู้ป่วย hemophilia A ซึ่งมี factor VIII inhibitor โดยเฉพาะการผ่าตัดที่เกี่ยวข้องกับกระดูกนั้นเป็นปัญหาใหญ่เนื่องจากมักจะมีปัญหาเลือดออกไม่หยุดแม้ว่าจะให้การรักษาด้วย factor VIII bypassing agent ที่มีใช้อยู่ก่อนที่จะมีการใช้ activated factor VII การใช้ activated factor VII เป็นการกระตุ้นให้เกิดการแข็งตัวของเลือดโดยใช้ extrinsic pathway โดย tissue factor ซึ่งพบว่าได้ผลเป็นที่น่าพอใจ รายงานต่อไปนี้เป็นประสบการณ์การผ่าตัด sequestrectomy ของ ilium ที่โรงพยาบาลจุฬาลงกรณ์

ประชากรศึกษา : ผู้ป่วยชาย อายุ 25 ปี ได้รับการวินิจฉัยว่าเป็น severe hemophilia A with chronic osteomyelitis of right ilium และมี factor VIII inhibitor 165 Bethesda unit ได้รับการรักษาแบบประคับประคองจนกระทั่งระดับ inhibitor ลดลงเหลือ 3.5 Bethesda unit ก่อนที่จะได้รับการผ่าตัด sequestrectomy of right ilium โดยใช้ recombinant activated factor VII (Novoseven®) 90 mg/kg IV push ก่อนผ่าตัดและ 16.5 mg/kg/h IV continuous infusion ระหว่างและหลังการผ่าตัด

ผลศึกษา : ผู้ป่วยเสียเลือดมากหลังจากเสร็จการผ่าตัดและใน 48 ชั่วโมงแรกต้องได้รับ pack red cell ทดแทน จนเกิด massive blood transfusion และเพิ่มขนาดยาขึ้นเรื่อยๆ จนเลือดหยุดหลังจากให้ 90 mg/kg IV push ทุก 2 ชั่วโมง เป็นเวลา 5 วัน ระดับ factor VIII inhibitor กลับสูงขึ้นเป็น 50 Bethesda unit ในวันที่ 5 หลังผ่าตัดเนื่องจากการให้เกร็ดเลือดเข้มข้นและพลาสมาทดแทนในระหว่างที่เกิด massive blood transfusion แต่ไม่ได้ทำให้เลือดออกมากขึ้น ค่า prothrombin time สั้นลงกว่า control ตั้งแต่วันแรกของการผ่าตัด ค่า APTT เริ่มสั้นลงเมื่อเทียบกับก่อนให้ยาในวันที่สองและเริ่มยาวขึ้นในวันที่ห้าหลังผ่าตัด

สรุป : การให้ activated factor VII สามารถช่วยให้ทำการผ่าตัดสำเร็จได้โดยไม่ต้องใช้การรักษาอื่นร่วมด้วย แต่การให้ขนาดยา 16.5 mg/kg/hr IV continuous infusion ระหว่างและ 2-3 วันแรกของการผ่าตัดไม่เพียงพอที่ทำให้ผู้ป่วยเสียเลือดเท่ากับคนปกติได้ ขนาดของยาที่สามารถควบคุมให้ผู้ป่วยไม่เสียเลือดมากกว่าปกติคือ 90 mg/kg IV push ทุก 2 ชั่วโมงใน 6 วันแรกของการผ่าตัด ไม่พบผลข้างเคียงของยาในผู้ป่วยรายนี้

Key Words: ● Sequestrectomy ● Ilium ● Hemophilia A ● Factor VIII inhibitor
● Recombinant activated factor VII

Glanzmann Thrombasthenia : Immunologic Diagnosis at Siriraj Hospital

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Platelet glycoprotein IIb/IIIa (GP IIb/IIIa) is a major platelet integrin with the composition of GP IIb and GP IIIa. The GP IIb molecule, designated CD41, consists of heavy and light chains. This CD41 antigen is expressed in both megakaryocytes and normal platelets. It is absent or defective on the platelets of patient with Glanzmann thrombasthenia. We studied the GP IIb/IIIa in 4 patients who were previously given diagnosis of Glanzmann thrombasthenia (3 patients) and unknown type of congenital platelet dysfunction in one patient on basis of the routine platelet function tests (bleeding time, platelet adhesion, platelet aggregation and clot retraction) by using the purified monoclonal mouse antibody for human CD 41. The methods of studies included the flow cytometry and the immunocytochemical study using Rapid Alkaline Phosphatase-labeled Streptavidin Biotin (AP-LSAB) kit. The results of the studies revealed very small amount (almost absence) of the GP IIb/IIIa on platelet of all 4 patients, as opposed to normal amount in their parents. These immunological technics of study for the presence of GP IIb/IIIa would be helpful for and also confirm the diagnosis of Glanzmann thrombasthenia.

Key Words: ● Glanzmann thrombasthenia ● GP IIb/IIIa

Intravenous Gammaglobulin for Treatment of Chronic Idiopathic Thrombocytopenic Purpura in Children

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Seventeen children, aged 3-16 years with chronic idiopathic thrombocytopenic purpura were given intravenous gammaglobulin. The patients divided into 2 groups. The first group consisted of 10 patients with the age of 4-16 years (5F, 5M). Two of the 10 children had undergone splenectomy; the other 8 had not. They received intravenous intraglobin (Biotest Pharma GMBH, Germany) in a dose of 400 mg/kg per day for 5 consecutive days. This group of children were studied during June 1990 to April 1995. The second group consisted of 7 children with the age of 3-15 years (4F, 3M). Splenectomy was not done in this group of children. They received intravenous Venoglobulin (Alpha therapeutic Corporation, USA) in a dose of 1 g/kg per day for 2 consecutive days. The later group were studied during March 1996 to August 1997. Three of the 10 children had a good response to the 5 day course of gamma globulin therapy, and 7 had fair response. Two children whose spleen was removed had fair response. One booster dose of 400 mg/kg of Intraglobin was given I.V. to 6 children who had fair response, the platelet count was increased in all cases but less than $100 \times 10^9/L$. Splenectomy was done in 3 of 4 patients who had fair response about 9-10 months later; 2 patients responded very well to splenectomy. One booster dose of 400 mg/kg of Intraglobin was given I.V. to 3 children who had good response, the platelet count was increased in all cases with only one was greater than $100 \times 10^9/L$. All of the children had increased in serum Ig G following Intraglobin except one (who had undergone splenectomy). No serious side effect necessitating cessation therapy. The most common side effect observed was nausea & vomiting (4), headache (3) and rise in temperature (1). Six of 7 children had good response to the 2 day course of gammaglobulin therapy, and one case had fair response. One booster dose of 1 g/kg of Venoglobulin was given I.V. to patient who had fair response, the platelet count was uncreased but less than $100 \times 10^9/L$. Four children who had good response were given one booster dose of 1 g/kg of Venoglobulin, the platelet count was increased in all children with only one case was greater than $100 \times 10^9/L$. All of the children had increased in serum IgG following Venoglobulin with no serious side effect. The most common side effect observed was rise in temperature(6), headache(5) and nausea & vomiting(2).

Key Words: ● Intravenous gammaglobulin ● Chronic idiopathic thrombocytopenic purpura

Splenectomy without Platelet Transfusion for Adult Autoimmune Thrombocytopenic Purpura (ATP) : Response, Relapse-free Survival and Predictive factors

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Study objective : To determine the result, complication and long term response of splenectomy without platelet transfusion for adult ATP patients and to establish predictive factors for complete response

Design : Retrospective-prospective descriptive study

Setting : Single center, Songklanagarind university hospital

Patients and methods : 48 adult patients with ATP undergone splenectomy without platelet transfusion during 1983-1996. Results of splenectomy were classified as complete response (CR, platelet $\geq 150 \times 10^9/l$), partial response (PR, platelet between $50-150 \times 10^9/l$) and no response (NR, platelet $< 50 \times 10^9/l$ or increment $< 25 \times 10^9/l$). Eight variables were analyzed as predictive factors for CR : sex, age duration from first diagnosis, duration of steroid administration, response to steroid, dosage of steroid before splenectomy, preoperative platelet count, time required for platelet response $> 150 \times 10^9/l$.

Results : There were 38 females and 10 males. Age ranged 15 to 64 years with mean age of 33 years. Median duration from first diagnosis was 8 months (range 0.5-106) with median duration of steroid administration of 16 weeks (range 1-240). Before splenectomy, patients received steroid with the median dosage of 45 mg/d (range 5-100). Elective surgery was performed in 40 patients (83%) and emergency surgery in 8 patients (17%). For elective cases, 22 was steroid-dependent and 18 was steroid-resistant. Indications for emergency splenectomy were intracranial hemorrhage in 6 (intracerebral 4, subarachnoid 2), acute appendicitis in 1, and concurrent severe autoimmune hemolytic anemia in 1. Platelet count before splenectomy ranged $2-404 \times 10^9/l$ with the median of $45 \times 10^9/l$. Median estimated blood loss during operation was 150 ml (range 30-700) and accessory spleen was found in 15%. No bleeding complication occurred. Only one patient developed postoperative subphrenic abscess. Platelet count rose to $> 150 \times 10^9/l$ within the first 3 postoperative days in 26 patients (62%). Overall response rate was 89% (CR 66%, PR 23%). With the median follow-up time of 60 months, the relapse-free of CR patients were 86% at 3 years and 74% at 5 years. Later 3 NR and 1 PR patients died because of intracranial bleeding. Univariate analysis indicated that age ≤ 35 years, daily dosage of steroid ≤ 40 mg, steroid respondent, and platelet response within the first 3 days were associated with CR, but only platelet response within 3 days retained predictive influence in multiple regression model. No postsplenectomy overwhelming infection was demonstrated.

Conclusions : Splenectomy can be performed safely in ATP patients without platelet transfusion. Platelet response within 3 days is the only significant predictive value for CR. The response rate and long term relapse-free rate were impressive

Key Words: ● Splenectomy ● Platelet transfusion ● Autoimmune thrombocytopenic purpura

Flow Cytometric Analysis of P-glycoprotein (P-gp), and Glutathione S-transferase-pl (GST-pp) in Monoclonal Gammopathy of Undetermined Significance, Plasmacytoma, Multiple Myeloma, and Amyloidosis

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Multiple myeloma is not a curable disease. Most of the patients relapse after plateau phase. Even high dose chemotherapy cannot eradicate the malignant cells from bone marrow. Drug resistance is a major problem in spite of effective chemotherapy in this disease. We studied drug resistance markers, P-glycoprotein (P-gp) or P-170 and glutathione S-transferase-pl (GST- π) in a total of 43 patients with monoclonal gammopathy, which included eight cases with monoclonal gammopathy of undetermined significance (MGUS), five with plasmacytoma (PCM), nineteen with multiple myeloma (MM: six newly diagnosed, seven plateau, five refractory, one relapse) and eleven amyloidosis (seven newly diagnosed, four after treatment). Using 3-color flow cytometry, a plasma cell gate was selected on the basis of 38+/45 - staining and the population was examined for the expression of the MDR marker. Data collected show that P-gp and GST- π were positive in MGUS, PCM and in all stages of MM and amyloidosis except P-gp could be negative in MM (1/19). These data suggest there are mechanisms of resistance in plasma cell dysclasia other than P-gp and GST- π . Relative resistance due to P-gp and GST- π mechanism is likely to be intrinsic property of plasma cells in myeloma and may provide a partial explanation why these diseases always relapse.

Key Words: ● P-glycoprotein ● Glutathione S-transferase-pl ● Monoclonal gammopathy of undetermined significance ● Plasmacytoma ● Multiple myeloma ● amyloidosis

Enhanced detection of P-glycoprotein Using a Commercial Available Lysing Solution

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Multidrug resistance (MDR) is a clinical problem in treatment of human malignancy. MDR1 gene and its product, P-glycoprotein (P-gp) are thought to play a major role. One of the most popular techniques use to study P-gp is flow cytometry. The expression of P-gp in malignant cells is sometimes very weak and hard to interpret. We used MRK16, UIC2 and C219 monoclonal antibodies (MoAbs) to detect P-gp in KG-1A, U266, 8226/DOX6 and 8226/DOX40 cell lines with or without fixation. We demonstrated that a method using a commercial brand lysing solution (FACS brand lysing solution: Becto Dickinson, San Jose, CA) as a fixative could enhance the detection of P-gp by flow cytometry. We used the MRK16 and the UIC2 MoAbs each of which detects an external epitope. The results show that the lysing solution enhanced detection of P-gp expression 2-4.2 fold with the UIC2 MoAb and 1.6-3.2 fold with the MRK16 MoAb.

Key Words: ● P-glycoprotein ● Multidrug resistance

Bcr-abl Gene Arrangement in Chronic Myelogenous Leukemia (CML): Clinical and Prognostic Significances - Preliminary Report

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Objectives : To characterize the type of bcr-abl gene rearrangement in patients with CML and to determine the relationship of breakpoint position to clinical manifestations, prognosis and survival

Design : Prospective descriptive study

Setting : Single center, Songklanagarind university hospital

Patients and methods : 31 adult patients (age ≥ 15 years) with CML were analysed for clinical and hematological data at diagnosis, Sokal prognostic index, duration of chronic phase and survival. The reverse transcriptase-polymerase chain reaction (RT-PCR) and nested PCR were used to detect the location of bcr-abl gene rearrangement. Cytogenetic study for Philadelphia chromosome was also performed in all of the patients.

Results : The frequencies of b3a2 (3' breakpoint) and b2a2 (5' breakpoint) rearrangement in patients with CML were 61.3% and 38.7%, respectively. The clinical features (age, sex preference, duration of symptoms before diagnosis, % hepatomegaly, liver size, % splenomegaly, and splenic size), hematological data (hemoglobin level, white blood cell count, platelet count, % basophils, and % blasts) and Sokal prognostic indices (median, 1.29 vs 1.26) of patients presenting the b3a2 or the b2a2 arrangement was not significantly different. With the median follow up time of 14 months for patients with b3a2 transcript, there was again no influence on the median duration of chronic phase (>24 vs 31.9 months).

Conclusions : The frequencies of bcr-abl gene arrangement in our CML patients are similar to the literature. This preliminary report suggests that the type of transcript has no clinical association or prognostic significance for CML patients. The larger amount of patients and the longer duration of follow-up are required.

Key Words: ● Bcr-abl ● Chronic myelogenous leukemia

Immunophenotypic Studies of Acute Leukemia in Adult : A Study of 103 Thai Cases

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Background : Immunophenotyping is used widely to phenotype cases of acute leukemia, which is now clearly important in classification. The areas of greatest practical importance are in morphologically difficult or undifferentiated cases and aberration patterns of markers in both ALL and AML. At present, the data on immunophenotypic distribution of adult Thai patients with acute leukemia is meagre.

Objective : To define the phenotypic patterns in adult ALL and AML patients, using the combination of CD45 intensity with right-angle light scatter (SCC).

Study design : From February 1996 to November 1997, 103 peripheral blood and/or bone marrow samples obtained from acute leukemic patients (age ≥ 15) diagnosed at the Hematology Unit, Chulalongkorn Hospital were tested. Fourteen monoclonal antibodies were used as the panel. All cases were analysed by flow cytometer (FacScan®).

Results : Twenty-six patients (25%) were defined as ALL. Of these, 20 (77%) were Pre-B ALL and 6 were T-ALL (23%). Two cases of Pre-B ALL and five cases of T-ALL had myeloid coexpression (CD13-33). Seventy-seven patients (75%) were AML. Of these, 38 (49%) were M1, M2, 13 (17%), M3, 14 (18%), M4, 9 (11%), M5 and 3 (4%), M7. Of the M1, M2 patients, four cases revealed CD19 coexpression and seven had CD7 biphenotype.

Conclusion : By using flow cytometry, 75% of adult acute leukemic patients were AML and 25% were ALL. Pre-B ALL contributed 77% of ALL. AML, M1, M2 accounted for 49% of AML cases. Interestingly, 17% (18 cases) of the patients were classified as biphenotypic leukemia.

Key Words: ● Immunophenotype ● ALL ● ANLL

The Association of Epstein-Barr Virus and Non-Hodgkin's lymphoma in Thai Patients

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The association of Epstein-Barr virus (EBV) and non-Hodgkin's lymphomas (NHLs) has been reported in most parts of the world. In this communication, we study the presence of EBV in 51 NHLs patients diagnosed in Chulalongkorn Hospital, Bangkok, Thailand. Immunohistochemical stainings for EBV latent membrane protein-1 (LMP-1) and phenotyping are done on paraffin sections. Forty cases (78.4%) are B-cell NHLs, 9 (17.6%) are T-Cell NHLs and 2 (3.9%) are unphenotyped. EBV is found in 16 tumors (30.77%); 13 in B-cell NHLs (31.7% of B-cell lymphomas), 2 in T-cell NHLs (22.2% of T-cell lymphomas), and 1 in unphenotyped NHLs (50% of unphenotyped tumors). Of 40 B-cell NHLs, the EBV + tumors are 8 of 25 diffuse large cell lymphomas, 2 of 4 mantle cell lymphomas, 2 of 3 follicular lymphomas, and 1 of 2 Burkitt's lymphomas. The rest of B-cell tumors; 1 mixed cell type, 3 medium-cell type, 1 Burkitt's like, and 1 MALT lymphoma, are all EBV -. Of 9 T-cell NHLs, the EBV + tumors are 1 of 4 peripheral T-cell lymphoma (PTCL) large cell type and 1 of 2 PTCL medium cell type. The rest which are 1 enteropathy associated T-cell lymphoma (EATCL), 1 PTCL, mixed cell type, and 1 lymphoblastic lymphoma, are all EBV -. Concerning the site of involvement, 12 tumors are nodal NHLs, 3 are B-cell + EBV + and 2 are T-cell + EBV +. One tumor in base of tongue, 2 palate, and 2 in buccal mucosa are all B-cell + EBV +. Four tumors are in tonsils; all are B-cell + and EBV +. Five cases are gastric lymphomas and only 1 is EBV +. Two tumors in thyroid gland are B-cell + EBV +. One tumor in spinal cord is EBV +. Seven tumors in soft tissue, 2 in brain, two involving skin, in kidney, two tumors in nasal cavity, 1 each in pharynx, nasopharynx, duodenum, caecum, anal canal, liver, and heart are all EBV-. The result suggests that EBV is involved in oncogenesis of non-Hodgkin's lymphomas and may be related with type and primary site of tumor.

Key Words: ● Epstein-Barr virus ● Non-Hodgkin's lymphoma

A Predictive Model of Life-Threatening Febrile Neutropenia After CHOP Chemotherapy in Aggressive Non-Hodgkin's Lymphoma

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Due to its curability and high incidence, non-Hodgkin's lymphoma, intermediate (aggressive) grade, is an important malignant disease in Thailand. Following the standard chemotherapy regimen, some patients however develop life threatening complication i.e, febrile neutropenia. The objective of this study is to identify the factors related to the occurrence of febrile neutropenia in aggressive NHL patients after standard CHOP chemotherapy. Eighty one cases of newly diagnosed aggressive NHL treated with CHOP were included. Pretreatment evaluation for disease status, performance status, staging of disease, and blood tested for complete blood count, serum creatinine, LDH, LFT and bone marrow biopsy were performed. By follow-up CBC at the time of nadir count or at the occurrence of fever, twenty one patients (25.9%) were found to have febrile neutropenia ($BT \geq 38.5^{\circ}C$, $ANC \leq 0.5 \times 10^9/L$). By using stepwise logistic regression, pre-treatment serum total bilirubin, serum albumin and bone marrow involvement ($p = 0.0007$, 0.0012 and 0.0010 , respectively) were strongly associated with the developing of febrile neutropenia. In conclusion, serum bilirubin, serum albumin, and bone marrow involvement are factors that significantly correlated with the occurrence of febrile neutropenia in aggressive NHL patients post first course CHOP chemotherapy. Patients with these clinical features might benefit from primary G-CSF prophylaxis in order to ameliorate this life-threatening complication.

Key Words: ● Febrile neutropenia ● CHOP ● Non-Hodgkin's lymphoma

Low dose rhG-CSF for Prophylaxis of Chemotherapy Induced Neutropenia in non-Hodgkin's Lymphoma

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Background : In the treatment of non-Hodgkin's lymphoma, the use of standard dose granulocyte-colony stimulating factor (rhG-CSF) at 5 mcg/kg/day for chemotherapy induced neutropenia has shown strong evidence of clinical benefits. However, the efficacy of low-dose rhG-CSF in non-Hodgkin's lymphoma patients undergoing standard CHOP chemotherapy has not been evaluated.

Method : This was a prospective, randomized trial of low dose granulocyte-colony stimulating factor (lenograstim at 50 mcg) in intermediate grade non-Hodgkin's lymphoma patients with severe chemotherapy-induced neutropenia. Patients were evaluated for efficacy of low-dose therapy on the prevention and recovery from neutropenia.

Results : Patients were randomized to receive 50 mcg of rhG-CSF (12 patients) and 100 mcg of rhG-CSF (16 patients). The two groups had similar backgrounds. The nadir neutrophil counts were significantly higher ($p < 0.05$) compared to observation period for the 100 mcg group. The duration of neutropenic days was also significantly reduced for the both groups.

Conclusion : Low dose rhG-CSF is very effective in improving chemotherapy induced neutropenia in intermediate-grade non-Hodgkin's lymphoma patients. This trial has shown that low dose therapy may deliver cost-effective therapy without compromising standard care.

Key Words: ● G-CSF ● Neutropenia ● Non-Hodgkin's lymphoma

Allogeneic Bone Marrow Transplantation in Osteopetrosis Patient : A First Case Report in Thailand

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We reported 7 year old who was diagnosed with osteopetrosis since 1990. His skeletal survey, peripheral blood picture, and bone marrow examination were consistent with osteopetrosis. We performed HLA-matched sibling (one year and ten month old brother) allogeneic bone marrow transplantation. The conditioning regimen included busulfan 16 mg/kg and cyclophosphamide 200 mg/kg. Graft versus host disease prophylaxis (GVHD) included cyclosporin A and methotrexate. Engraftment, GVHD, and chimerism information and other details during and after bone marrow transplantation will be discussed.

Key Words: ● Bone marrow transplantation ● Osteopetrosis ● GVHD

Therapeutic Outcome and Prognostic Features of Adult Patients Undergoing Bone Marrow and Peripheral Blood Stem Cell Transplantation at Chulalongkorn Hospital

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Objectives : To determine outcome and prognostic features of adult patients treated with BMT or PBSCT at Chulalongkorn Hospital.

Study Design : The data were retrospective reviewed on the patient's demographic features, treatment option, complications and survival and relapse - free survival.

Statistical Analysis : Survival rate was analysed by Kaplan - Meier analysis. Prognostic features were determined by log - rank (univariate) and Cox - proportional hazard (multivariate).

Results : A total of 80 patients were treated from April 1990 - December 1997. The median age was 33 year (range, 15-55). Male : female was 1.1:1. Thirty patients had acute myelogenous leukemia, 20, NHL, 13, aplastic anemia, 9, CML, 5, breast cancer, 2, ALL and 1, myeloma. Seventy-two percent of the patients were classified as low-risk (aplastic anemia, AML in 1st CR, CML chronic phase, NHL 1st CR, breast cancer with ≥ 6 positive axillary nodes and myeloma) and 27% as high-risk (AML not in 1st CR, NHL, not in 1st CR, CML, accelerated or blast phase, ALL and breast cancer with metastasis). Thirty five patients (43.5%) underwent HLA-identical allogeneic transplant (29, bone marrow, 6 peripheral blood), 43, autologous PBPC and 2 syngeneic PBPC. Chemoradiotherapy (TBI, endoxan) was the main preparative regimen given in 40% of the patients. The mean CD 34 positive cell dose in patients treated with autologous PBPC was 5.0×10^6 /kg (range, 0.12-34). All patients showed hematopoietic engraftment. Twenty-four percent of patients undergoing allogeneic transplant developed acute GVHD (3 with grade III, IV). Three patients had chronic GVHD. At time of analysis, the 5 year overall survival rate was 60%. At a median follow-up time of 9.0 months, 14 patients (17%) relapsed. By using stepwise Cox proportional hazard, risk group was the only factor significantly associated with relapse (RR = 7.0, 95% CI = 2.4 - 25.2) and death (RR = 2.6, 95% CI = 1.1 - 6.0). The 5-year overall survival of patients with low-and high-risk were 72% and 30% respectively (p=0.007).

Conclusion : The long-term overall and relapse-free survival of patients treated with BMT and PBSCT at Chulalongkorn Hospital was 60%. The most significant predictor of the outcome was the risk-group of the patients. The treatment option had no impact on the outcome of the patients.

Key Words: ● Bone marrow transplantation ● Peripheral blood stem cell transplantation

Quality of life of Thai Patients Post Bone Marrow and Peripheral Blood Stem Cell Transplantation

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Backgrounds : Quality of life (QOL) has recently emerged as an important tool used for measuring therapeutic efficacy in clinical studies, particularly in the realm where medical intervention leads to survival benefit but with an apparent risk of side effects and functional impairment such as bone marrow transplantation (BMT). Whereas a number of study has recently been paid on the QOL of patients undergoing bone marrow and peripheral blood stem cell transplantation (PBSCT) in the western population, data on the post-transplant QOL in Asian patients is sparse.

Objectives : To determine the QOL of Thai patients undergoing BMT/PBSCT and compare the data with those reported from the western countries. The variables that had impact on the patients' QOL were determined.

Settings : Two tertiary-care university hospitals.

Population : Patients age ≥ 15 years undergoing BMT/PBSCT from April 1991 to July 1997 at Chulalongkorn Hospital and from February 1991 to October 1995 at Ramathibodi Hospital who were in disease-remission were entered into the study.

Measurements : The QOL questionnaire, modelled after several previous instruments used for assessment QOL in BMT survivors, were sent by mail to the patients. The questionnaire consists of two instruments: the demographic data tool for determining patients' characteristics and the Quality of Life-BMT survivor (QOL-BMT) tool. The QOL-BMT tool included 37 items representing the four domains of QOL, i.e., physical status and abilities, psychological well-being, social interactions and treatment-related symptoms. The items are scaled on 5 point Likert-type scales that "1" equals "worst outcome" and 5 equals "best outcome." Participants filled out the questionnaires based on their experience within the last two weeks and returned by mail. A randomly selected sample of patients who had completed the initial QOL survey were asked to repeat this survey two weeks later for the test-retest analysis.

Results : There were a total of 62 patients. The median age of the patients was 35.5 years (range, 16-56). Male:female was 1:1. The median time from transplant was 9.1 months (range, 1-62.8). Eighty-nine percent of the patients had malignant diseases. While 30 patients had allogeneic transplant, 31 were treated in the

autologous setting. Thirty-five patients had peripheral blood as the stem cell source. High-dose chemoradiotherapy was given as the preparative regimen in 58% of the patients. Reliability of the questionnaire measured as the test-retest (Pearson's correlation) and the internal consistency (Cronbach's α coefficient) were 0.79 and 0.87 respectively which indicated strong stability. The mean score of the overall QOL of the patients was 3.76 on a scale of 1-5 with "5" being their best. The mean scores of the physical domain, psychological domain, social domain and treatment-related well-being were 3.94, 3.99, 4.12 and 4.20 respectively. By using the univariate analysis, the following variables were found to have statistical significant association with the patients' overall QOL : depression, despair, anxiety/tension, happiness, satisfaction, memory, appearance, family acceptance, financial burden, fatigue, dyspnea, functional ability (ECOG), able to attend work/school, and the length of time since BMT/PBPCT. None of the variables on the socioeconomic status nor the sexual functioning had a statistical significant association with the patients' QOL. However stepwise multiple regression revealed only four independent variables that had statistical significance on the QOL namely, despair, appearance, functional ability and financial burden. These variables accounted for 53% of the variance in the overall QOL scores.

Conclusion : The mean score of the overall QOL of Thai patients post BMT/PBSCT was 3.76 (scale of 1-5) which was comparable to most other studies from western countries. The four most important variables that had impact on the patients QOL were despair, appearance, functional ability and financial burden. The association of financial burden and QOL had not formerly been noted.

Key Words: ● Bone marrow transplantation ● Peripheral blood stem cell transplantation