66 Audiologic Assessment for β-Thalassemia Major Patients with Long-term Transfusion Therapy
接受長期輸血的乙型地中海貧血患者之聽力檢查

Kang-Hsi Wu[1], Yu-Hua Chao[2,3], Ching-Yu Lin[4], Ming-Hsui Tsai[4], Ching-Tien Peng[1], Chia-Der Lin[4]
Department of Pediatrics, China Medical University Hospital[1]; Department of Pediatrics, Chung Shan Medical University Hospital[2]; School of Medicine, Chung Shan Medical University[3]; Department of Otorhinolaryngology-Head and Neck Surgery, China Medical University Hospital[4], Taichung, Taiwan
巫康熙[1]、趙玉華[2,3]、林菁俞[4]、蔡銘修[4]、彭慶添[1]、林嘉德[4]
中國醫藥大學附設醫院兒童醫學中心[1]；中山醫學大學附設醫院兒童部[2]；中山醫學大學醫學系[3]；中國醫藥大學附設醫院耳鼻喉部[4]

Background: The combination of life-long RBC transfusion therapy with chelating agents is a treatment choice for patients with β-thalassemia major. Some investigators have proposed auditory impairment related to the use of deferoxamine. However, the mechanisms remain unclear and whether or not deferiprone has similar side effects needs to be evaluated.
Methods: Thirty-seven patients with β-thalassemia major who received regular transfusion in our hospital were enrolled. Chelation agents, including deferoxamine and deferiprone, were used. To assess audiologic function, otoscopy, pure tone audiometry (PTA), tympanometry, transient evoked oto-acoustic emission (TEOAE), and auditory brainstem response (ABR) were conducted. Bithermal caloric test was performed to evaluate vestibular function.
Results: All of the 37 patients had normal findings on otoscopic evaluation and their tympanograms were type A. Thirteen patients (35.1%) had hearing impairment at one or more frequencies as detected by PTA. Compared to those without hearing impairment, patients with hearing impairment had lower serum ferritin levels (p=0.01). Seven of 21 patients (33.3%) failed to pass the TEOAE, while 13 (61.9%) had abnormal ABR findings. Sixteen patients (80%) had canal paresis in the caloric test.
Conclusions: The incidence of auditory impairment and vestibular dysfunction are high in patients with β-thalassemia major, and potential lesions involved in auditory impairment may exist anywhere along the auditory pathway. Hearing impairment is significantly more common in patients with lower serum ferritin levels. Thus, regular check-ups of serum ferritin levels and periodic audiologic assessment are mandatory.

67 Lower Factor XII level in Patients with Polytransfused Beta Thalassemia Major
重型地中海貧血病患表現較低的第十二凝血因子

Te-Fu Weng[1], Kang-Hsi Wu[1], Ching-Tien Peng[1,2], Ming-Ching Shen[3,4]
Department of Pediatrics, China Medical University Hospital[1]; Department of Biotechnology and Bioinformatics, Asia University[2]; National Taiwan University Hospital[3]; Changhua Christian Hospital[4]
翁德甫[1]、巫康熙[1]、彭慶添[1,2]、沈銘鏡[3,4]
中國醫藥大學附設醫院[1]；亞洲大學生物科技學系[2]；國立台灣大學醫學院附設醫院[3]；彰化基督教醫院[4]

Background: The patients with beta thalassemia major received poly-transfusion have many changes on the coagulopathy. The previous studies postulated that damage of tissue from iron overload resulted in a fall in the contrast factors. It further postulated that iron overload would lead to lower plasma level of factor XII. Therefore we performed a comprehensive evaluation of contact activation pathway and ferritin level in such patients.
Methods: 24 patients (mean 22-years-old, range from 12 to 37 years-old) affected by beta-thalassemia major were studied. All patients received hyper-transfusion program and took regular oral iron chelation therapy with Deferiprone. The average annual volume of blood transfusion, factor XII level and ferritin level were recorded.
Results: We found that factor XII of these patients were significantly lower. (Mean 42.6%, range from 22.7 to 71.2) But there is no significant correlation of patients' age, volume of blood transfusion, ferritin level. There is also no association with factor XII level to cardiac iron.
Conclusions: In our data, there could not be confirmed that iron overload associated with lower level of factor XII. The causes of factor XII decline in poly-transfused beta-thalassemia major patients need further investigation.

68 Single Institution Experience of Unrelated Cord Blood Transplantation for Primary Immunodeficiency
非親屬臍帶血移植治療原發性免疫不全症候群：單一機構之經驗

Tang-Her Jaing[1], Wei-I Lee[2], Tsung-Yun Ch... Shih-Hsiang Chen[1], Chao-Ping Yung[1], Iou-Jih Hung[1]
Division of Hematology/Oncology[1], Allergy, Immun... and Rheumatology[2], Department of Pediatrics, Chang... Children's Hospital, Chang Gung University, L... Taoyuan
江東和[1]、李文益[2]、張從彥[1]、陳世翔[1]、楊兆平[1]、... 林口長庚紀念醫院兒童內科部血液腫瘤科[1]、風濕... 疫科[2]

Background: Pediatric patients with ... immunodeficiencies (PID) constitute medical emer... In the absence of an HLA-identical hematopoietic s... donor, unrelated donor cord blood transplantation ... another treatment option. There is little data r...