which was diagnosed by brain MRA and/or cerebral angiography in Taipei Veterans General Hospital since 1990 till 2012 were retrospectively reviewed. Cases with moyamoya syndrome and brain tumors were enrolled for study.

Results: Within total 98 patients with moyamoya vasculopathy, there are 10 boys and 3 girls who had brain tumors, including 7 craniopharyngiomas, 5 optic pathway tumors (4 of them were associated with neurofibromatosis type 1) and 1 dysembryoplastic neuroepthelial tumor (DNET). All of the tumors except DNET are located in sellar or suprasellar region which is in vicinity to circle of Willi. The DNET is located over right anterior mesial temporal region, which is also in vicinity to circle of Willi. Six of the 13 patients had moyamoya syndrome without radiation exposure, and the median duration from diagnosis of brain tumor to diagnosis of moyamoya syndrome was about 3 years and 7 months. For the other 7 patients who had radiation exposure, the median period for developing moyamoya syndrome after radiation was 7 years (range from 7 months to 10 years and 7 months).

Conclusions: The common feature in our patients with brain tumor who developed moyamoya syndrome is the location of the tumor, which is in vicinity of circle of Willi. Radiation does not seem to be a triggering factor for moyamoya syndrome in this retrospective study.

111 Tourette Syndrome Increases Subsequent Depression Risk: A Taiwanese Population-based Cohort Study

妥瑞症候群會增加隨後抑鬱症的風險：臺灣群體之世代研究

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Background: A possible association exists between Tourette syndrome (TS) and subsequent depression. This study evaluates the relationship between TS and depression in Taiwan.

Methods: Claims data from the Taiwan National Health Insurance database were used to conduct retrospective cohort analyses. The study cohort contained 1337 TS patients who were frequency matched by sex, age, urbanization of residence area, parental occupation, and baseline year with 10 individuals without TS. Cox's proportional hazard regression analysis was conducted to estimate the effects of TS on depression risk.

Results: In patients with TS, the risk of developing depression was significantly higher than in patients without TS (p-value for log-rank test <.0001). After adjusting for potential confounding, the TS cohort were 4.85 times more likely to develop depression than the control cohort (HR=4.85, 95%, CI=3.46-6.79).

Conclusions: In Taiwan, patients with TS have a higher risk of developing depression. The findings of this study are compatible with studies from other countries. This study could provide an evidence to inform the prognosis for a child with TS. The mechanism between TS and increased depression risk requires further investigation.

112 Becker Muscular Dystrophy Patients with Point Mutations of Dystrophin Gene

肌縮蛋白基因點突變之貝克氏肌失養症患者發生早期心臟病變之現象

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Background: Becker muscular dystrophy (BMD) is an X-linked muscular dystrophy caused by the dystrophin gene mutations, allelic to Duchenne muscular dystrophy. Cardiac involvement (CI), as one of clinical features of BMD patients, usually can be subclinical during adolescence and early adult life. However, the severity of CI has poor correlation to the degree of skeletal muscle weakness, often leading to late identification. We thus attempt to identify early predictors of CI in BMD patients for early intervention.

Methods: We analyzed the BMD patients with regular follow-up in our hospital from August, 1990 to May, 2012. All patients were diagnosed by clinical features and patchy pattern of dystrophin immunostaining from biopsied muscles. The clinical features, serum creatine kinase (CK), body mass index, mutations of the dystrophin gene, electrocardiography (ECG), echocardiogram, and spirometry were recorded and analyzed. Total 12 BMD patients were recruited with age distribution of 10 to 52 years (Mean=26; SD=12) and then divided into two groups for further analysis based on the genotypes (deletion vs. point mutation, n=5 vs. 7).

Results: Between these two genotypic groups, there is no significant difference in terms of current age, age of onset, age of showing positive Gowers` sign, age of non-ambulation, serum CK and BMI. In patients with point mutations, 4 of 7(57%) had abnormal echocardiogram findings and 6 of 7(85%) had abnormal ECG findings. In patients with deletions/duplications, 2 of 5(40%) had abnormal ECG findings and no patient had abnormal echocardiogram findings.

Conclusions: In BMD patients, symptoms of CI were very mild or asymptomatic. Delay of treatment may cause heart failure or sudden death. In our study, the proportion of CI is exclusively high in BMD patients with point mutations. The BMD patient with point mutations need early and regular cardiac evaluation.