Mortality

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Mortality Problems of Congenital Adrenal Hyperplasia in Central Java-Indonesia: 12 years experiences

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Introduction: Congenital Adrenal Hyperplasia (CAH) is an autosoma recessive inheritance due to the defect in the pathway that converts cholesterol into cortisol leading to decreased production of cortisol. Patients with CAH need life long treatment of glucocorticoid and sometimes mineralocorticoid. Early recognition and treatment of CAH can prevent adrenal crises. However, in our cohort of the classic type of CAH, the late-identified patients with CAH are more prevalent than early-identified patients who are different with countries that already have CAH newborn screening. Moreover, fatal adrenal crises are seen in clinical practice. Reports on mortality in patients with CAH in Indonesia are lacking. The aims of this present study were to report and identify the mortality problems of CAH in Indonesia.

Methods: A retrospective study was done in the Center for Biomedical Research (CEBIOR) Semarang, Central Java. Data related to mortality was derived from research database from 2004-2016.

Result: Of 78 patients with CAH (74 females and four males) in our cohort database, ten patients (12, 8%) were reported died due to adrenal crises. Among these ten patients had died of adrenal crises, six patients with CAH have already confirmed with hormonal and mutation analysis and four patients confirmed base on clinical and hormonal analysis. The mean age of diagnosis was 8.4 months (ranged from 1 month to 5.5 years). The average age of death was 22.3 month (ranged from 2 months to 6 years).

Among 78 patients, only 5% are 46 XY CAH, which may indicate that adrenal crises in male newborn might be under-diagnosed or unidentified. Nine siblings of seven families had a history of siblings who died because of a probable salt wasting condition before diagnosis confirmation. Thus the mortality rate of CAH in Indonesia is probably higher than this rate.

Conclusion: There was a high mortality rate of CAH in Central Java, Indonesia due adrenal crises. The high mortality rate can be anticipated or reduced if the Hydrocortisone and Fludrocortisone were available and affordable for patients with CAH. Findings from this study are strongly recommend for promoting early diagnosis and treatment as well as improving compliance of young patients with CAH. In addition, awareness of the medical community in the rural area and endorsement to the policy maker for availability of medication are essential to decrease the mortality rate of patients with CAH in Indonesia.

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