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PROPHYLACTIC THYROIDECTOMIES IN MEN2 SYNDROME: MANAGEMENT AND OUTCOMES

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Aim of the Study: The purpose was to evaluate the outcomes of prophylactic thyroidectomies performed in an academic setting in the context of multiple endocrine neoplasia type 2 (MEN2) syndrome.

Methods: We reviewed charts of patients under 18 years old who underwent prophylactic thyroidectomy for a MEN2 syndrome at a free standing children's hospital between April 2006 and October 2015.

Main Results: The study included 21 patients (57% female) with a mean age of 6.2±2.5 years old at surgery. Nineteen patients had MEN2A syndrome with RET proto-oncogene mutation identified at genetic screening, the remaining two were RET-negative with familial medullary thyroid cancer (FMTC). One patient (C620R) had a concomitant Hirschsprung's disease. All patients were asymptomatic at first evaluation. Of the 11 patients who had RET proto-oncogene mutations ranked as Moderate Risk for medullary thyroid cancer (MTC) by the American Thyroid Association (7 with V804L, 2 with V804M, 1 with C620R, 1 with C618R), one (V804L) had a microcarcinoma on the resected specimen and the others had C-Cell Hyperplasia. Among the 8 patients who had RET proto-oncogene mutation ranked as High Risk level for MTC (4 with C634R, 4 with C634F), all patients had microcarcinoma. Of the nine patients with microcarcinoma, three underwent surgery after 5 years of age. No microcarcinoma exceeded 6 mm. There were no permanent complications. Six patients experienced transient hypocalcemia of which only one was symptomatic. No patients had lymph node involvement and no recurrence was noted during the follow-up period (mean duration 4.1±3.4 years).

Conclusions: Of 19 children with MEN2A syndrome who underwent a prophylactic thyroidectomy, nine had undetected microcarcinoma. This study highlights the need for a complete familial history including FMTC history and a preventive surgical approach.