Medullary thyroid cancer (MTC) is a unique form of thyroid cancer and distinct from the more common differentiated variants (papillary and follicular). As a neuroendocrine tumor, MTC is derived from the parafollicular cells, or C cells, of the thyroid; the secretion of calcitonin and carcinoembryonic antigen (CEA) enables these serum peptides to be utilized as pre- and postoperative tumor markers, which correlate with the extent of disease and tumor burden. This is in contrast to papillary and follicular thyroid cancer where the use of thyroglobulin as a measure of disease extent is much less sensitive and specific. Serum levels of calcitonin and CEA can be utilized to guide the extent of preoperative imaging, the results of which influence the extent of initial surgery. MTC which is localized to the neck remains largely a surgical disease as there is no effective adjuvant therapy currently available to minimize disease recurrence. In addition, while the majority of patients have sporadic disease, approximately one-fourth of patients with newly diagnosed MTC will have an inherited form, secondary to multiple endocrine neoplasia (MEN) type 2A or 2B, or the related syndrome of familial MTC.

These unique biologic features can make the management of patients with MTC quite challenging and require the multidisciplinary expertise of internists, pediatricians, endocrinologists, genetic counselors, medical oncologists, pathologists, and thyroid surgeons. This text is not meant to replace the current American Thyroid Association guidelines, last published in 2015, which serves as the point of reference for the management of patients with MTC; but rather, we hope to provide a more in-depth discussion of some of the clinical challenges which physicians and patients face.

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