



## Abstract

**Objectives:** to describe for the first time epidemiological characteristics and clinical features of medullary thyroid carcinoma at tertiary care center in Costa Rica.

**Materials and methods:** we reviewed the Thyroid Clinic records for the last 5 years and selected all those cases with a diagnosis of medullary thyroid carcinoma. Statistical analysis was performed using SPSS 20.0

**Results:** 23 patients were identified and 22 records were retrieved and analyzed. We were unable to locate 1 of the records. There were 16 women and 6 men. Mean age was 51.09 years. Diagnosis was made on average at 42.36 years. One case was diagnosed in the setting of MEN2A and another one in MEN2B. These 2 cases were the only ones with family history of MEN. 10 cases (45.5%) presented initially as thyroid nodule, 31.8% as neck mass, 18.1% as goiter (multinodular) and only one case as submandibular lymph node. 64.5% of patients had some kind of neck dissection as initial surgery. 22.7% required a second surgery, usually neck dissection or completion thyroidectomy. Initial histology showed multifocality in 2 cases, 5 cases with vascular invasion, 2 with perineural invasion, 4 with extracapsular invasion. 45.4% had lymph node metastases on initial presentation. 2 patients had distant metastases. TNM staging showed stage I in 22.7%, II 4.5%, III 36.4%, IVA 4.5% and IVC 9.1%. 31.8% had recurrence. After initial surgery, none of the patients had an undetectable calcitonin level. Regarding therapy, 3 patients received radiotherapy and one patient TKI. After a mean follow up of 7.8 years, 1 patient died after 4 years. 4 (18.1%) have no evidence of clinical or biochemical disease (with undetectable calcitonin level). 16 patients have stable disease (5 with distant metastases) and one patient is pending surgery. 3 of the patients had concurrent papillary thyroid carcinoma.

**Conclusion:** This is the first description of a case series of medullary thyroid carcinoma in Costa Rica. It is more prevalent in women and in most cases not related to MEN. Most patients have a total thyroidectomy plus neck dissection and have stable disease.

## Introduction

Medullary thyroid cancer (MTC) typically accounts for 3-4% of all thyroid cancers. The clinical course of MTC can be indolent, remaining unchanged for years, while some cases can be aggressive, associated with high mortality.

Autosomal dominant inherited diseases is present in 25% of cases, caused by a germ line mutation in the rearranged during transfection (RET) proto-oncogen; typically multifocal or bilateral. Sporadic forms occurs in 75% of cases, typically unifocal.

3% of all thyroid cancers are MTC, it affects 1 in 30000 people. Age at diagnosis is older for sporadic (51 years) than genetic (21 years).

Since there are no published data of medullary thyroid carcinoma in Central America, epidemiological data is unknown.

The main objective of the present study is to describe epidemiological characteristics of medullary thyroid carcinoma at a tertiary care center in San José, Costa Rica.

## Materials and Methods

San Juan de Dios Hospital is a tertiary care center located in San José, Costa Rica. Our Thyroid Clinic is multidisciplinary team that includes endocrinologists, pathologist, neck surgeons, and nuclear medicine specialists. All thyroid cancer cases are discussed in this Clinic.

We reviewed the Thyroid Clinic records at the San Juan de Dios Hospital in Costa Rica for the last 5 years (January 2010-December 2015) and selected all those cases with a diagnosis of medullary thyroid carcinoma. Statistical analysis was performed using SPSS 20.0.

## Results

23 patients were identified and 22 records were retrieved and analyzed. We were unable to locate 1 of the records. There were 16 women and 6 men. Mean age was 51.09 years. Diagnosis was made on average at 42.36 years.

Figure 1. Clinical presentation of MTC.

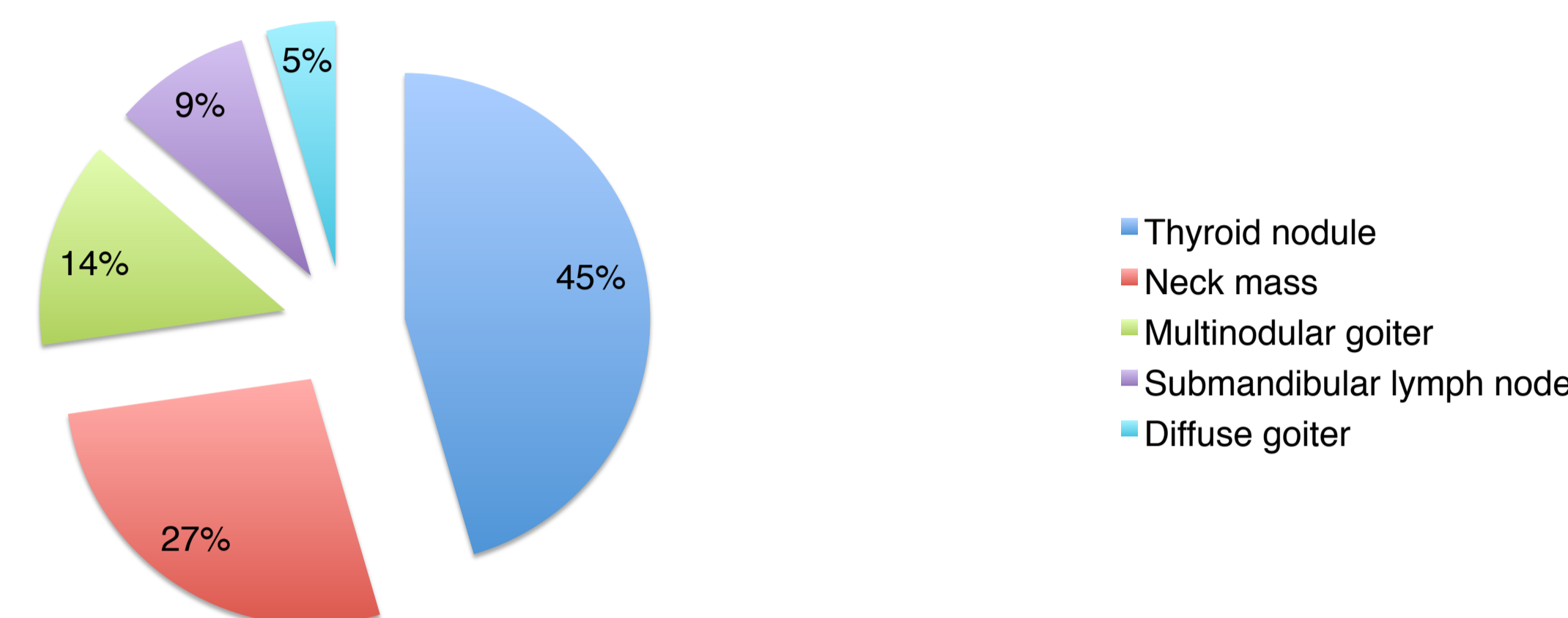


Figure 2. TNM staging in the initial presentation with medullary thyroid carcinoma (%)

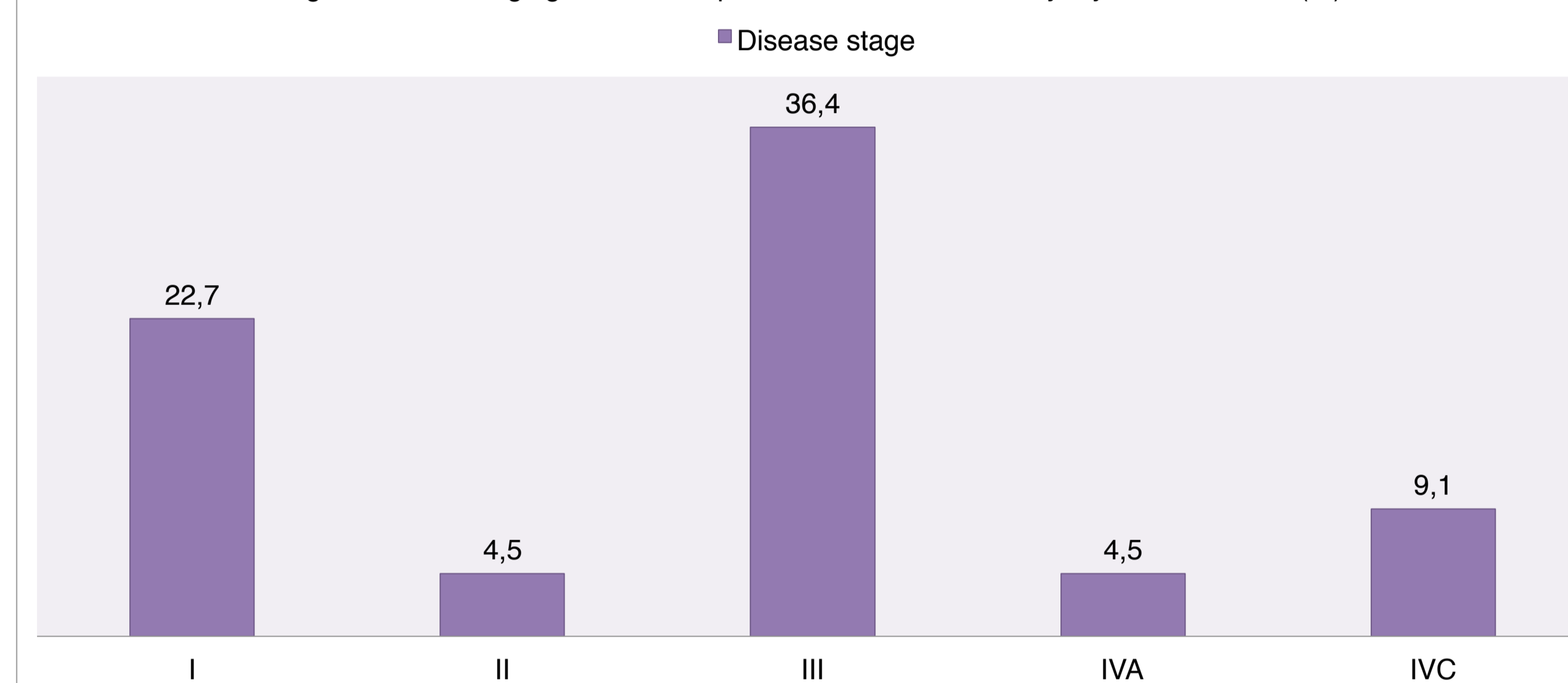


Table 1. Initial histologic characteristics

Characteristic	N	%
Multifocality	2	9.5
Vascular invasion	5	23,8
Perineural invasion	2	9,5
Extracapsular invasion	4	19
Lymph node metastases	9	42,9
Distant metastases	2	9,5

One case was diagnosed in the setting of MEN2A and another one in MEN2B. These 2 cases were the only ones with family history of MEN. 64.5% of patients had some kind of neck dissection as initial surgery. 22.7% required a second surgery, usually neck dissection or completion thyroidectomy. Regarding therapy, 3 patients received radiotherapy and one patient TKI.

After a mean follow up of 7.8 years, 1 patient died after 4 years. 4 (18.1%) have no evidence of clinical or biochemical disease (with undetectable calcitonin level). 16 patients have stable disease (5 with distant metastases) and one patient is pending surgery. 3 of the patients had concurrent papillary thyroid carcinoma.

Figure 3. Mean calcitonin levels depending on latest clinical evaluation

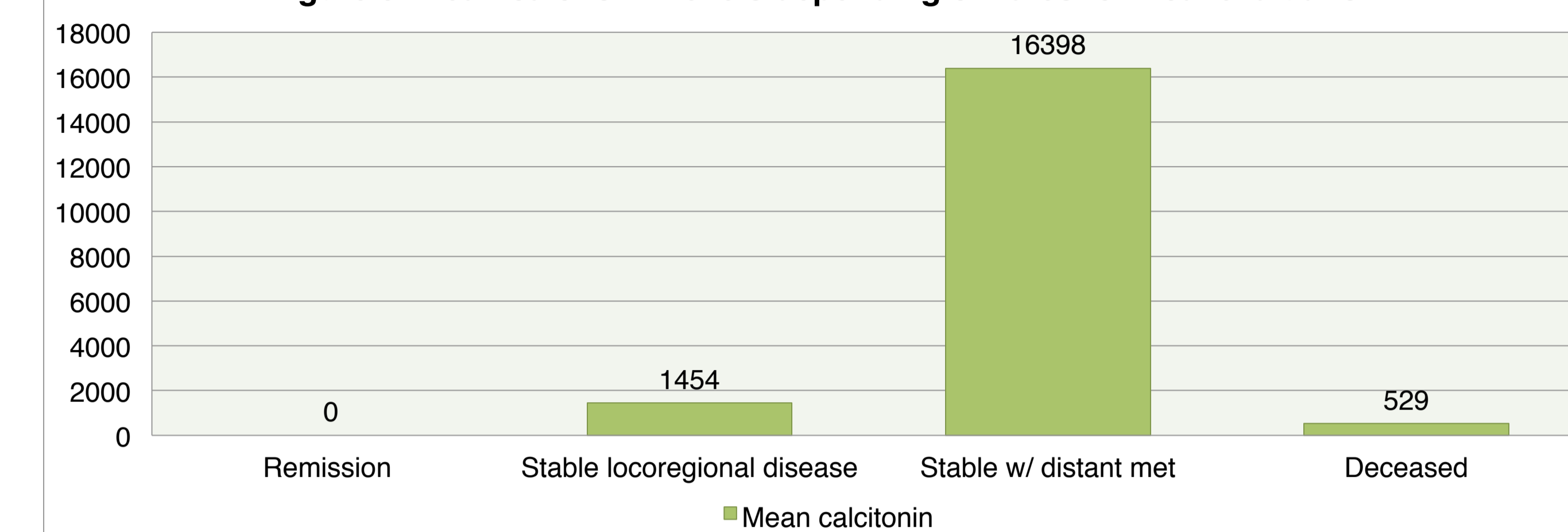
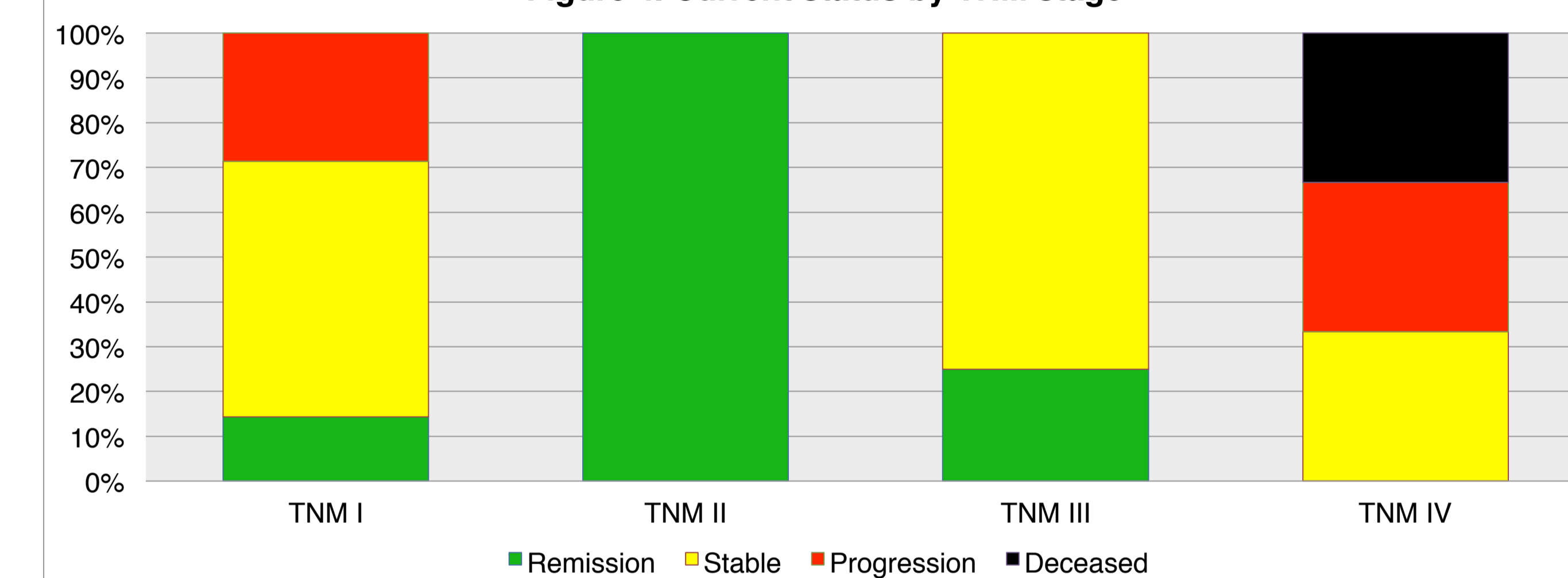


Figure 4. Current status by TNM stage



## Discussion

During 2010-2015 a total of 540 cases of new thyroid carcinomas were diagnosed at the Hospital San Juan de Dios, 23 of which were medullary thyroid carcinoma. It represents 4.25% of all thyroid cancers, which is in line with other series(3-10%). Our data showed an incidence of 0.4 cases per 100000 persons per year. Mean age in our series is 51.06 years and is within what has been reported, usually between 4<sup>th</sup> and 6<sup>th</sup> decades of life.

In recent times, there has been no significant trend toward an earlier stage of disease at diagnosis, as just under half of the patients present with stage III or IV. Similar to that, our series showed that 50% of our patients are diagnosed in stage III or IV, although TNM staging was unknown in 19% of patients.

After a mean follow up of 7.8 years, only 1 patient died and he had a T4 disease at presentation. The clinical behavior of sporadic MTC is unpredictable and some patients with distant metastases may live for several years. The ten years survival rates for patients with stage I, II, III and IV is 100%, 93%, 71% and 21%. 18.1% respectively. 4 (18.1%) of our patients have no evidence of clinical or biochemical disease (with undetectable calcitonin levels), 31.8% had recurrence. 16 patients have stable disease, including 5 with distant metastases.

## Conclusions

This is the first description of a case series of medullary thyroid carcinoma in Costa Rica. It is more prevalent in women and in most cases not related to MEN. Most patients have a total thyroidectomy plus neck dissection and have stable disease. Clinical characteristics are similar to other series.

## References

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