

**THE RELATIONSHIP OF PAPILLARY THYROID CANCER (PTC)  
AND FAMILIAL ADENOMATOUS POLYPOSIS (FAP)**

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**QUESTIONS:**

1. Is there an association between PTC and FAP?
2. Is there an increased incidence of PTC in patients with FAP?
3. Should all patients with FAP be screened for the presence of PTC?

**CLINICAL ASPECTS**

A 14-year-old female underwent a colectomy with an ileo-anal anastomosis for FAP (Figure 1). At the age of 20, during a routine physical examination, she was found to have an incidental asymptomatic 1.5 cm right thyroid nodule. Fine-needle aspiration was highly suggestive of a PTC “with an unusual cytologic pattern.” A total thyroidectomy was performed for a bilateral, multicentric, node-negative PTC (Figure 2).

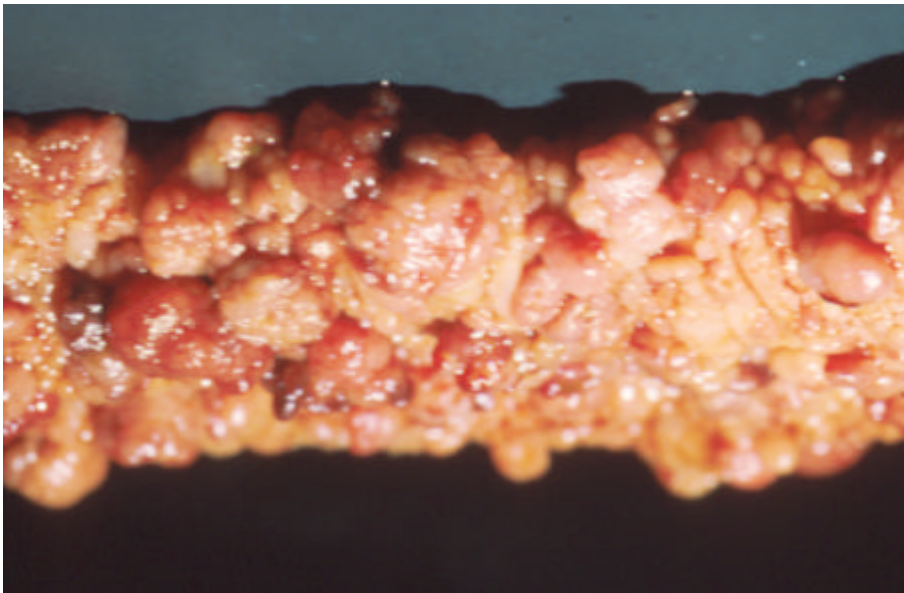


Fig. 1: Colectomy specimen with the classic appearance of multiple colonic polyps.



Fig. 2: Total thyroidectomy specimen demonstrating metastatic bilateral papillary thyroid carcinoma.

Histology demonstrated a papillary thyroid carcinoma with areas of papillary formation (Figure 3A), a “Swiss cheese” appearance (Figure 3B), and morulae formation with intranuclear “holes” (Figure 3C).

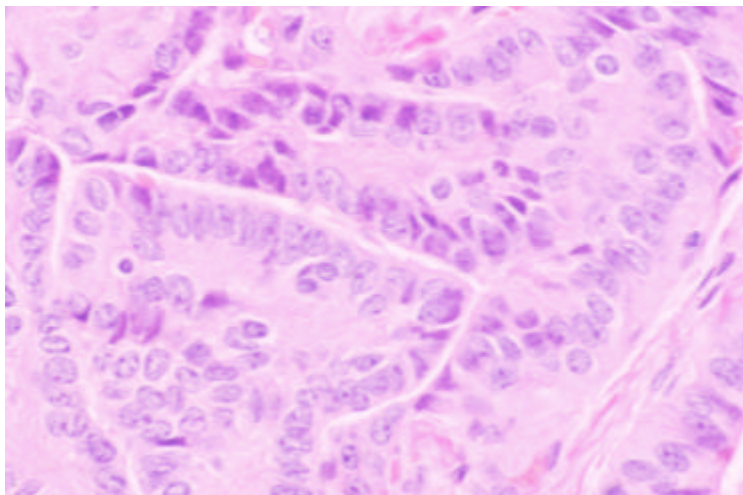


Fig. 3A: Histological appearance of papillary carcinoma with papillary formation.

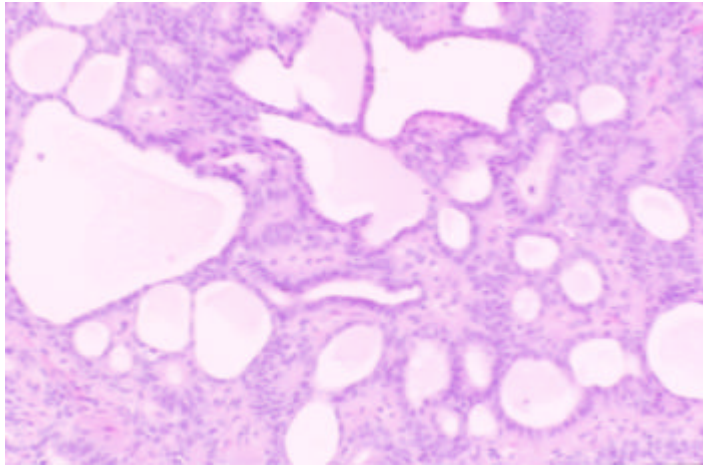


Fig. 3B: Histological appearance of papillary carcinoma with Swiss cheese appearance.

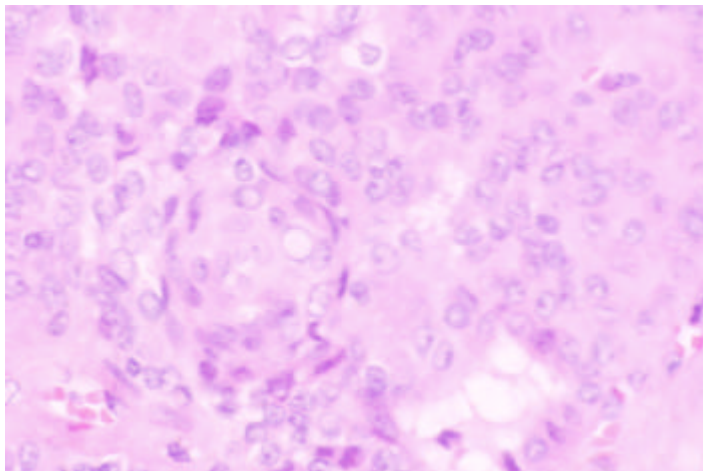


Fig. 3C: Histological appearance of papillary carcinoma with morulae formation.

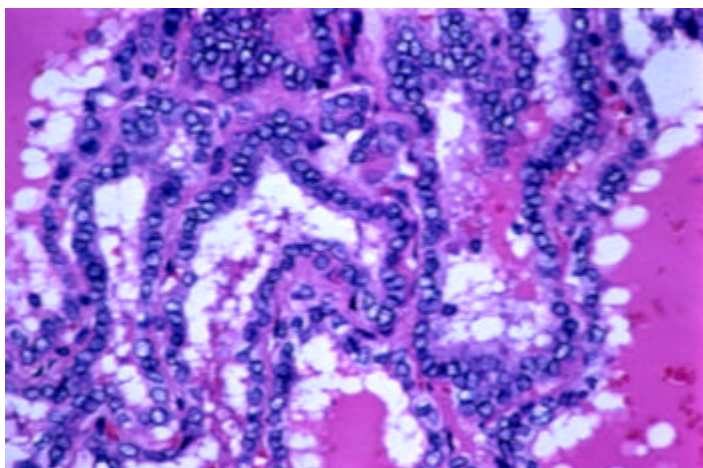


Fig. 3D: Histological appearance of type ordinaire PTC

The patient remains disease-free two years postoperatively.

## DATA SUMMARY:

The Welsh pathologist, Sir Delwyn Williams, should be given credit for not only bringing attention to this rare association, but for recognizing that the histological appearance of PTC occurring in patients with FAP is so unique and distinctive that the diagnosis of FAP could, in fact, be suggested based on the histology encountered in the thyroid malignancy! In addition to the unique histological pattern, the PTC in such patients differs from PTC encountered sporadically in that multicentricity, bilaterality, and regional nodal spread occurs in approximately 60%, 40%, and 20% of patients, respectively. This is in comparison to 30%, 80%, and 30% in patients with type ordinaire PTC.

There would appear to be a marked increase in the incidence of PTC in patients with FAP. Studies from England and the United States of American documented that the incidence of sporadic PTC was 2.4 and 4.6 per 1000,000 of the general population, respectively. The incidence for patients with FAP climbed markedly to 150 per 1000,000 in both of these studies (Figure 4).

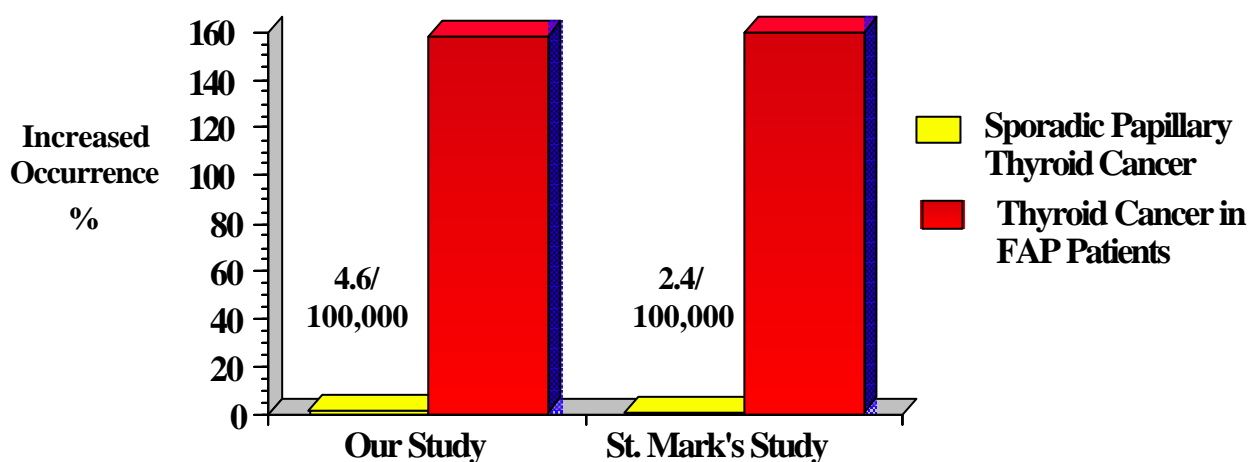


Fig. 4: Incidence of papillary thyroid carcinoma in familial adenomatous polyposis versus sporadic patients.

This increased incidence mandates routine screening (physical examination coupled with ultrasonography and liberal use of fine-needle aspiration when indicated) in all patients with FAP. Conversely, any patient with a PTC with unusual histopathology should be screened for FAP.

Once the diagnosis is confirmed, the PTC should be treated by total thyroidectomy and routine nodal sampling/dissection with adjuvant therapy ( $^{131}\text{I}$  and TSH suppression) as indicated.

Despite seemingly aggressive pathology, patients with FAP associated PTC follow a very favorable long-term course, as do most patients with sporadic PTC.

#### **ANSWERS:**

The response to the three initial questions is in the affirmative for all. There is a clear, unique, albeit rare, association between PTC and FAP. The incidence of PTC is increased markedly in FAP patients, mandating that all FAP patients be screened for PTC by physical examination, thyroid ultrasonography, and liberal use of fine-needle aspiration biopsy when indicated

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“Kind words can be short and easy to speak,  
but their echoes are truly endless.”

Mother Teresa