



Patient Report

The PJS/Thyroid Connection

A Report by
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This report is on the PJS/thyroid connection. For new readers, I've written several pieces on PJS from a patient's view. Some topics include: sex cord tumors, bowel preparations, and a review of the article *Very High Risk of Cancer in Familial PJS*⁷ etc. Please remember these pieces are not medical advice, but reviews of the literature that you can take to your doctor. Some of you may want to have your thyroids screened; others may have thyroid troubles that weren't connected to PJS by doctors.

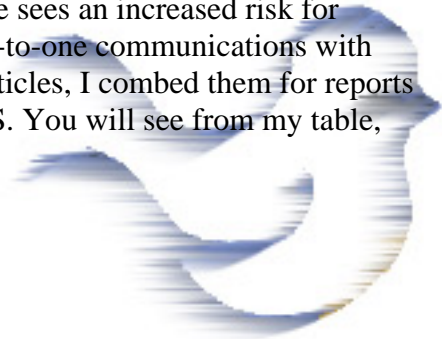
This topic was introduced last year by two of our support group members who visited Dr. Stratakis at the NIH in Bethesda, Maryland. Dr. Stratakis is an endocrinologist who works primarily on Carney Complex. His study¹ is open to people with PJS. One member wrote that her thyroid tumors were found during her study participation. Another member wrote to us on September 4, 2000:

"The doctor gave me a couple of his articles. His latest, from *Frontiers in Bioscience*, discusses the overlap between disorders associated with multiple tumors and pigmentation defects. Among the non- GI neoplasms associated with PJS, endocrine tumors including thyroid cancer and ovarian and Sertoli cell tumors are the most frequent. My surgeon in RI was unaware of any thyroid association with PJS. When I needed bloodwork for my surgery in May, I asked to get my thyroid level checked. All came back normal. But, Dr. Stratakis told me that it is not sufficient to just get bloodwork. You also need a thyroid ultrasound. He said that there is an especially aggressive form of thyroid cancer associated with PJS."

The full article from *Frontiers in Bioscience* is available online².

The news of potential thyroid cancer and screening for people with PJS was particularly startling since none of the screening guidelines from other doctors include the thyroid.³ I read Dr. Stratakis' article which included the following quote: "Among the nongastrointestinal neoplasms associated with PJS, endocrine tumors including thyroid cancer and ovarian and Sertoli cell tumors are the most frequent." He referenced three articles⁴⁻⁶, but when I read the articles, I found only one mention of thyroid cancer or tumors. I wrote to Dr. Stratakis requesting more information, but he didn't respond.

Dr. Giardiello in his article, *Very High Risk of Cancer In Familial PJS*⁷, showed no increased risk of thyroid cancer in PJS and doesn't even include it in his tables. I have to wonder what Dr. Stratakis knows that we don't know. If he sees an increased risk for thyroid cancer, it might be as a result of his studies or one-to-one communications with other doctors. Now that I have a large collection of PJS articles, I combed them for reports of thyroid tumors, malignant & benign, in people with PJS. You will see from my table, that there are reports of thyroid tumors in PJS folks.



Author (publication date)	Gender, Age	Diagnosis
Spigelman (1989) ⁶	Female, 30	Thyroid cancer
Burdick & Prior (1981) ⁸	Female, 61	Thyroid colloid nodule & Hashimoto's thyroiditis
Yamamoto M (1992) ⁹	Female, 29	Thyroid papillary adenocarcinoma
Reed MW (1990) ¹⁰	n/a	Papillary carcinoma of the thyroid
Boardman LA (1998) ¹¹	Age 33	Thyroid cancer
Srivatsa, PJ (1994) ¹²	Female, 91	Grade 1 metastatic papillary cystic adenocarcinoma of thyroid with squamous metaplasia 2 months after first operation (patient was 91 years old & in good health at time of article)
Srivatsa, PJ (1994) ¹²	Female, 21	Graves' disease, subtotal thyroidectomy
Srivatsa, PJ (1994) ¹²	Female, 32	Colloid adenoma of thyroid with small area of papillary carcinoma
Dormandy, TL (1957) ¹³	Female, 60	Nodular goiter
Dormandy, TL (1957) ¹³	Male, 52	Nodular goiter

The most common type of thyroid cancer in PJS folks and the general population is papillary carcinoma. A search at Cancer Net revealed that thyroid papillary adenocarcinoma is a differentiated cancer that is highly treatable and curable.¹⁴ So Dr. Stratakis' comment on an aggressive thyroid cancer associated with PJS isn't substantiated from my readings of the literature.

Though I did find cases of PJS folks with thyroid problems, but I have no idea if these findings are significant. Since thyroid problems are fairly common in the general population, there may not be a connection to PJS. Hopefully we can get response from medical folks. I will also forward this to Dr. Stratakis and Dr. Giardiello, asking for their opinions.

If you or someone you know has thyroid problems that may or may not be connected to PJS, it will be interesting to hear your stories. I wish my findings were clearer, but maybe a response will come from more knowledgeable doctors about the PJS/thyroid connection.



¹*Definition of the Genotype and Clinical Phenotype of Primary Pigmented Nodular Adrenocortical Disease and its Associated Conditions (Carney Complex).*

http://clinicalstudies.info.nih.gov/detail/A_95-CH-0059.html

²Stratakis, Contsantine A. *From Genetics of Carney Complex and Related Familial Lentiginoses and Other Multiple Tumor Syndromes.*

<http://www.bioscience.org/2000/v5/d/stratak/fulltext.htm>

³Please search our list archives for screening guidelines. You need to be a member of the list to search the archives. Contact the webmaster or the author for assistance.

⁴Giardiello, F. M., Welsh, S. B., Hamilton, S. R., Offerhaus, G. J. A., Gittelsohn, A. M., Booker, S. V., Krush, A. J., Yardley, J. H., Luk, G. D. *Increased risk of cancer in the Peutz-Jeghers syndrome.* New Eng. J. Med. 316: 1511-1514, 1987.

PubMed ID : 3587280

⁵Hizawa, K.Iida, M., Matsumoto, T., Kohrogi, N., H. Kinoshita & T. Yao. *Cancer in Peutz-Jeghers Syndrome.* Cancer November 1, 1993, Volume 72, No. 9.

⁶Spigelman, A. D., Murday, V., Phillips, R. K. S. *Cancer and the Peutz-Jeghers syndrome.* Gut 30: 1588-1590, 1989.

PubMed ID : 2599445

⁷Giardiello, Francis, et al. *Very High Risk of Cancer in Familial Peutz-Jeghers Syndrome.* Gastroenterology 2000;119:1447-1453.

⁸Burdick, D.; Prior, J. T. *Peutz-Jeghers syndrome: a clinicopathologic study of a large family with a 27-year follow-up.* Cancer 50: 2139-2146, 1982.

PubMed ID : 7127254

⁹Yamamoto M, Hoshino H, Onizuka T, Ichikawa M, Kawakubo A, Hayakawa S. *Thyroid papillary adenocarcinoma in a woman with Peutz-Jeghers syndrome.* Intern Med 1992

Sep;31(9):1117-9

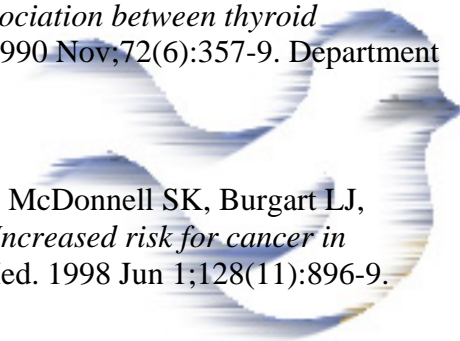
PMID: 1421720

¹⁰Reed MW, Harris SC, Quayle AR, Talbot CH. *The association between thyroid neoplasia and intestinal polyps.* Ann R Coll Surg Engl 1990 Nov;72(6):357-9. Department of Surgery, Royal Hallamshire Hospital, Sheffield.

PMID: 2241052

¹¹Boardman LA, Thibodeau SN, Schaid DJ, Lindor NM, McDonnell SK, Burgart LJ, Ahlquist DA, Podratz KC, Pittelkow M, Hartmann LC. *Increased risk for cancer in patients with the Peutz-Jeghers syndrome.* Ann Intern Med. 1998 Jun 1;128(11):896-9.

PMID: 9634427



¹²Srivatsa, PJ et al. *Disseminated Cervical Adenoma Malignum and Bilateral Ovarian Sex Cord Tumors with Annular Tubules Associated with Peutz-Jeghers syndrome.* Gynecological Oncology 53, 256-264 (1994).

¹³Dormandy, Thomas L. *Gastrointestinal Polyposis with Mucocutaneous Pigmentation (Peutz-Jeghers syndrome).* New England Journal of Medicine, 1957 June 13; 1141-1146.

¹⁴Cancernet for Thyroid Cancer. <http://cancernet.nci.nih.gov/pdqsrch.shtml>

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