Carney Complex (CNC)

Carney complex (CNC) is a rare genetic condition in which people typically have pigmented (dark) areas on their skin, as well as a higher risk of certain types of tumors.

- What are the effects of Carney complex (CNC)?
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- How often should people with Carney complex (CNC) be screened for tumors?
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What are the effects of Carney complex (CNC)?

In addition to having dark, pigmented areas on their skin, people with CNC are at higher risk for certain tumors including:

- **Endocrine tumors:** Tumors in organs or glands that make hormones, such as the pancreas, testicles, ovaries, or the adrenal, thyroid, parathyroid, or pituitary glands
- **Myxomas:** Benign (non-cancerous) tumors that develop most often in the heart (cardiac myxomas) but can also start in the skin or other organs
- **Schwannomas:** Tumors (usually non-cancerous) that start in Schwann cells, which normally wrap around and insulate nerves

Signs of CNC typically appear during ch ETn08ta3a othwhin aes,(oo1l)Tj 0 Ts 0 g ET Q q 0.79019h E

increase in number during puberty and may fade as people get older.

Cardiac myxomas are also common. While these tumors are non-cancerous (because they don't spread to other parts of the body), they can still cause serious health issues. These tumors can reduce blood flow in the heart and increase the risk of blood clots (which can lead to a stroke), and they can be life-threatening. Myxomas can also develop on the skin or in the mouth, breast, or other organs.

Endocrine tumors of the adrenal and pituitary glands and of the testicles (in males) are also common in people with CNC. Tumors in these glands or organs can result in abnormally high levels of specific hormones in the body:

- In adrenal gland tumors, high levels of cortisol can lead to weight gain, high blood pressure, diabetes, and easy bruising. This is known as **primary pigmented nodular adrenocortical disease (PPNAD)**.
  Pituitary gland tumors can lead to high levels of growth hormone, but this (known as **asymptomatic growth hormone hypersecretion**) usually doesn't cause any symptoms. Some people may developsgalyypersecretion.

children. For more information, talk with an assisted reproduction specialist at a fertility clinic.

How common is Carney complex (CNC)?

CNC is rare. Only several hundred cases have ever been reported worldwide.

How is Carney complex (CNC)diagnosed?

People are diagnosed with Carney complex based mainly on if they have certain criteria. CNC can be diagnosed:

- If you have 2 or more of the major diagnostic criteria below, OR
- If you are found to have a mutation in the *PRKAR1A* gene, OR
- If you have 1 major diagnostic criterion and a first-degree family member (parent, sibling, or child) with CNC

Major diagnostic criteria for CNC include:

- Spotty skin pigmentation
- Myxoma (of the heart, breast, skin, or mouth or other mucosal surface)
- Breast ductal adenoma (benign breast tumor)

- A yearly **echocardiogram** (ultrasound of the heart), beginning in infancy. People who have already had a cardiac myxoma should have an echocardiogram every 6 months.
- Yearly **endocrine (hormone) blood tests** such as cortisol, prolactin, and IGF-1, beginning in a person's teens.
- Yearly thyroid **ultrasound.**
- Yearly testicular ultrasound for boys, starting before puberty.

Screening recommendations may change over time as new technologies are developed and more is learned about Carney complex. It's important to talk with your health care team about appropriate screening tests.

How is Carney complex (CNC) managed?

Treatment of CNC is different for each person, depending on the type of tumor(s) they have, their symptoms, and other factors.

In general, most types of tumors resulting from Carney complex are removed with surgery if they are causing symptoms (or if they might cause problems in the future). Other types of treatments might also be helpful in some situations. Close follow-up is usually recommended after treatment to watch for possible signs of a tumor recurring (com59u34s?k) or new tumors form59u.

For some types of tumors that aren't likely to cause health issues, watching them closely (instead of removing them right away) might be an option.

Questions to ask the health care team

If you are concerned about your risk of Carney complex or the tumors it can cause, talk with your health care team. It might be helpful to bring someone along to your appointments to take notes. Consider asking your health care team the following questions:

- Could my family carry a gene mutation for Carney complex? How likely is it that I might have it?
- Should I meet with a genetic counselor or other genetics specialist for a risk assessment, and possibly genetic testing[3]? Can you refer me to someone?
- Should I get any screening tests to look for tumors? When should these start?

Hyperlinks

1. www.cancer.org/cancer/types/thyroid-cancer.html
2. www.cancer.org/cancer/types/bone-cancer.html
3. www.cancer.org/cancer/risk-prevention/genetics/genetic-testing-for-cancer-risk.html

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