

## Nontraditional Inheritance: Genomic Imprinting in Glomus Body Tumors

The most striking finding in this report was that the familial form of this disorder *is transmitted by males only*, either directly or through unaffected females, but *is not an X-linked condition*. Interestingly, these tumors are very much like other familial tumors, in that when they are familial, they are of early onset, occur at multiple sites, and have more severe symptoms and arise at a younger age than nonfamilial tumors.

This recent review by van der Mey et al of patients with glomus tumors identified 69 patients, of whom 34 had no family history and 35 had a family history. Another 82 patients were found within the families of the familial cases. Overall, there was a female excess, but this was entirely in those patients without a family history. This was not an X-linked condition because there was male-to-male transmission. The number of affected males and females in the familial cases was the same. The most appealing explanation for

these observations is *genomic imprinting*, in which a condition expresses itself only when inherited from a parent of one sex. The maternally derived gene is apparently inactivated during oogenesis in the mother but can be reactivated during spermatogenesis in her male offspring. In the case of glomus tumors, the family history suggests that when the gene is inherited from the mother, the genetic information is somehow suppressed; but when inheritance is from the father, the genetic information allows expression of the tumors over time. These are slow growing, benign, single or multiple tumors. They are known as chemodectomas or nonchromaffin paragangliomas and are derived from the glomus body tissue. They are most often found in the carotid body but also sometimes in the adventitia of the carotid bifurcation, the glomus jugulare, or the vagal body. Genomic imprinting seems to be common among familial and congenital tumors.

van der Mey AGL, Maaswinkel-Mooy PD, Cornelisse CJ, et al. Genomic imprinting in hereditary glomus tumors: evidence for a new genetic theory. *Lancet* 1989; 2:1291-1294.

**Editor's Comment**—*The concept of genomic imprinting is important and exciting. It may explain patterns of inheritance that have not previously been easily understood. Reexamination of a large pedigree, looking for differences in expression when the disorder is inherited from the mother versus the father, presents a whole new way of looking at information and understanding mechanisms of genetic expression.*

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