

Review and Editor's Comment: X Inactivation of the X Chromosome Is Not Complete

A series of papers recently published present concurring data indicating that the inactivated X chromosome is not completely inactivated. There are now known at least 4 areas of Xp and 2 areas of Xq of the inactivated X chromosome that are active. Those genes known to be subject to X inactivation, and those known to escape X inactivation on the X chromosome, are depicted in Figure 1. One of the 2 areas on Xq encompasses the inactivation center, which is believed to be responsible for inactivation of 50% of the X chromosomes. Intriguingly, current thinking regarding this center is that this area is active on the inactive X chromosome but inactive on the active X chromosome. This area on the inactive X chromosome, known as the X inactive specific transcript (XIST), produces a transcript, whereas the same area on the active X does not. This area is an area for a candidate gene that could be involved in influencing the process of X inactivation.

The importance of this phenomena is considered further in the references listed below. The readers are encouraged to

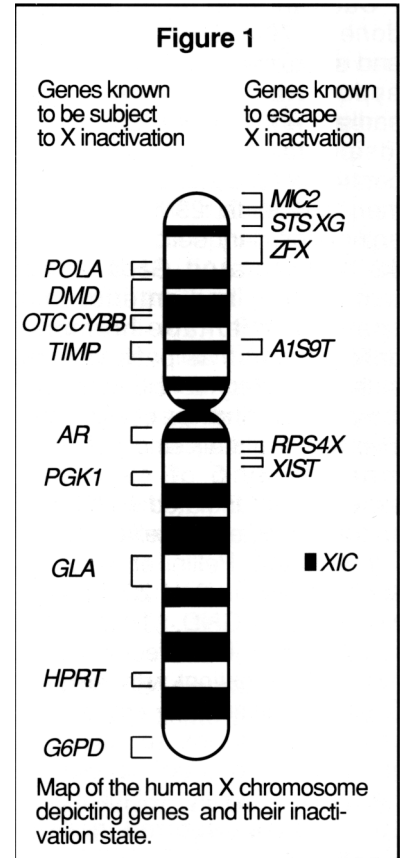
review these articles in detail, as Ohno's law of the constancy of the genetic material of the mammalian X chromosome is now being challenged by Watson et al after having been accepted for almost 25 years. The field is also intriguing as rare families manifesting female-to-female transmission of X-linked traits such as hemophilia B may have a co-inherited defect in the X inactivation center (XIC) resulting in the exclusive inactivation of the normal chromosome. XIST also may be involved in the phenotype of X-chromosome disorders such as Klinefelter and Turner syndromes. Deletion mapping in 46,XY Turner females has refined 1 probable Y chromosome localization to a 90 kb stretch between the possible sex determining gene, SRY, and the more proximal ZFY gene.

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