

The Tay Sachs Tie

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There are many cultural anomalies in South Louisiana, many things that are unique only to our community, such as Cajun and Creole cuisine, Zydeco music, and the *Courir de Mardi Gras*. These are just a few of the unique cultural attributes of South Louisiana. However, there is another, more sinister anomaly associated with our community—a disease called Tay Sachs. According to the March of Dimes website, in Louisiana, between 2021 and 2023, 416 infants died before reaching their first birthday, an infant mortality rate of 7.4 percent per 1,000 live births. According to the CDC, significant racial disparities exist. In 2022, the mortality rate for non-Hispanic Black infants was 11.9, more than double the 4.7 rate for non-Hispanic white infants. The possibility that Tay Sachs Disease contributes to the infant mortality problem also remains a feasible notion.

While most of us have never heard of Tay Sachs Disease, I believe it is a topic worth discussing, as it begs the question of how many of the infant deaths are related to the disease. We also must understand where the disease comes from. It is my contention that Tay Sachs awareness needs to be heightened in South Louisiana specifically. Furthermore, it is also my contention that Tay Sachs disease is even present at all in our population due to the frequent intermarriage between Louisiana natives and Jewish peoples and further that some Louisiana natives were in fact of Jewish decent. In the Jewish news publication *Forward*, in the article “Doctors Look to Raise Tay Sachs Awareness Among Louisiana Cajuns,” Nate Sugarman notes that

Tay-Sachs disease is caused by the congenital absence of a vital enzyme, hexosaminidase-A. Without this enzyme, the body cannot break down one of its fatty substances, which builds up abnormally in the brain and impairs the central nervous system. The gene that causes the infantile form of the disease is present in about one in 27 Ashkenazic Jews in the United States. About one in 250 Sephardic Jews and people of non-Jewish descent are also carriers. The disease that afflicts the Louisiana Cajuns is the same as the one that affects Ashkenazic Jews.

As members of this community, I think we owe it to ourselves as well as our children to be aware that we are susceptible to this disease.

I am not a doctor. Nor am I a scientist. However, the topic of Tay Sachs in the Cajun community became an interest of mine nearly fifteen years ago after the birth of my second child. Being Cajun myself, I was very concerned about the wellbeing of my baby. I have read and reviewed several articles on this subject during that fifteen-year space of time. Furthermore, it is an irrefutable fact that Cajun babies die from Tay Sachs disease. Yet it seems that, for the most part, Cajuns are unaware of the existence of this disease. This is evidenced by the sheer scarceness of information on the topic, and the obscurity of that which does exist. A research paper titled “The Presence of Two Different Infantile Tay-Sachs Disease Mutations in a Cajun Population,” which was printed in *The American Journal of Human Genetics*, explains that

within 3 decades, eight infants from six Cajun families in southwest Louisiana have been diagnosed with TSD. Of these, four apparently unrelated affected infants were born in the last 10 years. Although the carrier frequency of TSD in

the Cajun population of Louisiana is unknown, the occurrence of so many affected infants in a population of less than 1 million (Rushton 1979, p. 3; Smith 1990) suggests that it may be increased over that in the general population.

Furthermore, in an article in *Perspectives*, Hannah Meddaugh notes that “for many, the term ‘Cajun’ evokes romantic scenes of rural Louisiana life. In the world of genetic counseling, however, ‘Cajun’ represents a distinct and often overlooked American ethnic group.”

As a younger generation of Cajuns emerge, seemingly more well-educated than former generations, we should strive to raise awareness about Tay Sachs; however, there does not appear to be anyone advocating for said awareness. There are no existing coalitions or advocacy centers with regards to either Ushers or Tay Sachs, and only a handful of genetic counseling service providers. Once awareness occurs, a discussion of plausible solutions should ensue. While there is no known cure for the disease, there are preventative measures. A well-informed community and genetic counseling seem the most plausible solutions to the Tay Sachs problem. Even small attempts to share the concern about Tay Sachs would positively affect our community by improving awareness in small increments. For example, Ashkenazi Jews have greatly reduced incidences of the disease by being informed and utilizing genetic counseling. According to Sugarman, “Tay-Sachs disease has been significantly curbed in the Ashkenazic Jewish community, thanks to increased awareness and a comprehensive screening process—which is especially popular in Hasidic communities.” If the Jewish community can curtail Tay Sachs spread, so can Louisiana Cajuns.

A second consideration is the question of how or why Louisiana Cajuns are afflicted with a predominately Jewish disease. The most plausible explanation seems to be that our Cajun

ancestors and Jewish people had similar migration patterns. In the article “Globalization, Diasporas and Transnationalism: Jews in the Americas,” Judit Liwerant notes that

The Americas represent an exemplary case in which historical paths and changing conditions interact. Globality as a foundational element and globalization processes shaped the changes in the Americas’ social and communal structures, while the local and the global intertwined. Dynamic patterns of interaction developed historically between Jewish communities and their societal milieux, shaping Jews’ changing status as citizens in nation-states, holders of a transnational condition.

That being said, it is a known reality that we Cajuns originated from the southern parts of France and migrated first to Nova Scotia. After The Grande Derangement, we also migrated to Louisiana, which may offer a viable explanation for our having encountered Jewish people, who were traditionally merchants and traders. These facts are at least one explanation as to how Cajuns became carriers of Tay Sachs disease. However, it is a reality that many Jewish people also migrated to Louisiana throughout its history. What began with nomadic fur traders escalated to many well-known Jewish merchant families whose names many may recognize. A few examples of prominent Jewish names in South Louisiana which have become part of Cajun Culture are Meyers, Freedman, Schiffanni, Lemann, Godchaux, Newmon, Wormser, and Sternberg, and these are just a few of the more well-known names. Upon doing further research, it came to my attention that Jewish surnames frequent the family trees of Cajuns, names like Levi/Levy, Kerner, Kahn, Wolf, Frank, David, Abramson, Fortlouis, Felsenthal, to name a few. And while doctors and scientists attribute the anomalous Tay Sachs gene in our genetic pool to the bottleneck effect of the deportation of the Acadians and our subsequent geographical

isolation, it seems to me as if we might want to further pursue an historical inquiry into the intermarriage of the Acadian descendants and Jewish people, who are known to carry the gene.

Now, if you will, imagine our community as one where we are all aware of our susceptibility to Tay Sachs. To do so is to imagine we live in a community with a lower infant mortality rate, a community so well informed that it utilizes genetic counseling and curbs Tay Sachs instances substantially or even eradicates them. And proactively and intentionally educating Cajuns on this is still needed, since the disease which very few Cajuns still know about had been identified in 1990 in article titled “Genetics Down the Bayou.” Here, Hannah Meddaugh notes that

Additional rare autosomal recessive disorders have been identified in Louisiana Cajuns, attributed to Acadian founder effects. Two different recurrent HEXA mutations resulting in Tay-Sachs have been identified in Cajuns . . . In one rural community called Iowa (pronounced Eye-oh-way), the carrier frequency is higher than 1 in 10. Tay-Sachs is so prevalent in the region that it is known colloquially as “the Cajun disease.” Written records and oral history from early Cajun settlements describe “lazy baby” disease where apparently healthy infants would decline and die in early childhood; perhaps the legacy of undiagnosed Tay-Sachs.

It is important for me to note here that I was aware of Tay Sachs, which my grandmother used to refer to as “The Lazy Baby Disease.”

Still, this information and its import needs to be passed along by more formal means, using all resources available to my generation. We as a younger, likely more educated and potentially more well-informed generation should take the necessary steps of informing one

another about the disease, and if someone is at risk of passing the disease along, they can seek genetic counseling before they choose to procreate.

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