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CASE REPORT

Stereotactic radiosurgery does not appear to impact cochlear implant performance in patients with neurofibromatosis type II

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ABSTRACT

Introduction: Neurofibromatosis Type 2 (NF2) is a genetic condition associated with the presence of bilateral vestibular schwannoma and progressive sensorineural hearing loss. Treatment options include observation, stereotactic radiosurgery (SRS) or surgical resection. Historically, retrocochlear pathology was a contraindication to cochlear implantation (CI). However it is now recognized as viable, in an attempt to restore hearing and improve communication.

Thus, the aim of this study was to contrast auditory function in CI recipients with NF2 whose tumors were either observed versus initially treated with SRS.

Methods: A local review of hearing outcomes in 2 CI patients (3 ears total) with NF2 was undertaken and then pooled with the existing medical literature. Comparative post-implant outcome data, including pure-tone average and aided speech perception measurements was analyzed and compared between 8 subjects whose tumors were observed and 11 subjects who received pre-implantation SRS.

Results: Mean tumor size in the observation cohort was 0.81cm (.13cm to 1.50cm), and 2.34cm in the SRS group (0.10cm to 3.7cm). The mean reported duration of deafness was 22 months as compared to 71 months in the SRS cohort. Both groups demonstrated similar pre-implant candidacy criteria, average age and open-set speech recognition. Although disparate outcome measurements were utilized across studies, an analysis of post-implant open-set speech perception found no significant differences between groups.

Conclusion: Despite a small sample size and highly variable post implantation testing methods, patients who undergo SRS for NF2 associated Schwannoma prior to CI have similar hearing performance and benefit by having already completed definitive tumor management.

Keywords: neurofibromatosis, type, NF2, cochlear, implant, speech, outcomes, schwanomma

INTRODUCTION

Neurofibromatosis Type II (NF2) is an autosomal dominant disorder resulting from a mutation in a tumor suppressor gene (Merlin).[1,2] This disease is characterized by the growth of benign, bilateral vestibular schwanommas.[1,2] There are often associated central and peripheral nervous system tumors. Patients with this disorder are complex and challenge skull base surgeons in treatment planning for best possible outcomes.

Due to the location of these tumors, individuals with NF2 often exhibit progressive hearing loss, tinnitus, and vestibular disorders with possible facial nerve involvement.[1,2] Typical treatment for NF2 associated schwannomas includes observation, radiosurgery or tumor resection; with the ability to serially survey the site as requisite. However, irrespective of treatment, the prognosis is often bilateral profound hearing loss.[3] The spectrum of tumor effect can include multiple intracranial tumors with significant sensory deprivation, including vision loss.

As the auditory system degrades, hearing restorative surgery is considered. Historically Auditory Brainstem Implantation (ABI) presented the only opportunity. More recently cochlear implantation is considered in the presence of an anatomically preserved cochlear nerve. Literature supports improved hearing outcomes over ABI in addition to reduced surgical morbidity, and operative time.[1,2,3,4,5,6,7,8] The cochlear implant attempts to restore sound awareness and speech understanding by electrically stimulating the auditory nerve. However, the ability to successively survey a tumor site is compromised by the presence of the implant itself.[9] This becomes a major consideration in decisions surrounding tumor management and ultimate rehabilitation.

Factors impacting cochlear implant function in NF2 include; tumor management strategy, tumor size, duration of deafness, status of the contralateral ear, presence of other central tumors, global neural plasticity and age at implantation.[5]

Stereotactic Radiosurgery (SRS) is a treatment option employed to control tumor growth by applying focused beams of radiation to a target site with high conformity.[10] There is a breadth of supporting literature for management in sporadic schwannoma, however data is less robust for the management of patients with NF2.[10,11,12]

There is a developing impetus for early tumor control in an attempt to preserve the integrity of the auditory neural pathway. Growth in an observed tumor having undergone ipsilateral CI may necessitate treatment that would negatively impact hardware viability. Further, routine surveillance in an observed tumor with ipsilateral CI is problematic.

Individuals with NF2 comprise a relatively small subsection of those with profound hearing loss and there is little available data on post CI speech outcomes on patients undergoing observation or SRS. Thus, the objective of this study was to assess the potentially negative impact of SRS on audition and speech perception following cochlear implantation, by comparing outcomes to an observational cohort of cochlear implant recipients.

The question is substantive as upfront SRS reduces the need for future tumor management, but it is also important to appreciate the potential impact of radiation on audition. We report two patients who underwent SRS followed by CI and compare speech perception outcomes with a review of the literature on NF2 patient auditory function with disparate tumor management strategies.

MATERIALS AND METHODS

Following Research Ethics Board approval, a local retrospective review of patients with NF2 who underwent cochlear implantation following SRS at our facility was undertaken.

Additionally, a review of the literature was conducted in order to obtain post-implantation speech outcomes in patients with NF2 whose tumors were observed or received radiosurgery. A search of PubMed and Medline databases for all English-language articles, published between 2000 and 2015 generated 9 results relevant to this study. Studies met inclusion criteria if they examined speech perception outcomes in NF2 patients who had either undergone observation therapy or radiosurgery prior to implantation. Three publications that examined tumor resection as a pre-implantation treatment or failed to include post-implantation outcome measures were removed from consideration for this publication.

Post implantation audiometric data is compared between two groups; those tumors being serially observed and those who received SRS prior to implantation. Data collection for this study included patient demographics, age at implantation, gender, ear implanted, duration of deafness, tumor size, treatment and post-implant speech scores.

Due to the relatively few publications on this subject and the small sample size available, every effort was made to include only pertinent data sets for comparison. Speech perception tests used to measure performance outcomes included word recognition in quiet, the Consonant-Nucleus-Consonant (CNC) test, the City University of New York (CUNY) speech perception test, the Hearing In Noise Test (HINT) and Arizona State University's AZ-Bio Sentence Test .[13,14,15,16] These validated speech perception tests have traditionally been employed post-implantation to objectively measure changes in speech understanding at the consonant, word and sentence level. Individuals missing comparable outcome data in previous studies were removed from the current analysis. Further, studies that included patients undergoing tumor resection prior to cochlear implantation were also excluded.

Eight patients were observed, while 11 patients and 12 ears received SRS followed by CI. Post-implant speech perception outcomes were summarized in a table format (Table 2). Statistical analysis of analogous data was completed where possible, with *P* values <0.05 considered significant. No retrospective power calculation was possible. Individual missing data was indicated with a dash.

RESULTS

Case Study 1

A 38 year old female presented with a gradual, progressive sensorineural hearing loss in her right ear and several years of left profound loss. The patient denied any balance issues and reported no history of hearing loss in her family. She had no previous experience using any form of amplification. Initial audiometric findings indicated a flat, moderately severe sensorineural hearing in the right ear and a profound sensorineural hearing loss in the left ear. Unaided word recognition was 16% in the right and 0% in the left ear. Speech perception for sentences was 2% in the right ear (Az-Bio test).[16] Imaging illustrated bilateral vestibular schwannoma in addition to bilateral trigeminal schwannoma (Table 1).

The patient underwent staged Gamma Knife stereotactic radiosurgery (Leksell Gamma Knife Perfexion, Elekta, Stockholm) for both tumors with 12.5 Gy to the 50% isodose line. The patient reported no negative treatment effects. Bilateral tumor size marginally decreased for both lesions (Table 1).

She subsequently had simultaneous bilateral CI due to concerns regarding duration of deafness in both ears. Surgery was complicated by the presence of bilateral cochlear fibrosis. Full electrode insertion was successful, however required stylet for insertion. Post-implant auditory thresholds were within the normal range of hearing bilaterally, with a pure tone average of 18.3dB HL in the right ear and 20dB HL in the left ear. Twelve months post-implant speech scores are summarized in Table 2.

Case Study 2

A 77 year-old male presented with gradually progressive left sensorineural hearing loss. The right ear had been nonfunctional for many decades. The patient reported a history of balance issues and the presence of tinnitus. The patient used conventional amplification in the left ear since 1978, but had never worn amplification in his right ear. Initial audiometric findings indi-

Table 1. Case study participant demographic information and pre-operative speech perception results.

	Case Study – Participant Demographics											
	Sex	Age	Ear Implanted	Tumor Size (cm)	DOD (mos)	Contralateral Ear	Treatment	Vestibular Issues	Tinnitus	Previous Hearing Aid Use	Pre-Op WRS	Pre-Op AZ-BIO
Current Study	M	77	Left	0.10	240	Profound SNHL	SRS	Yes	Yes	Yes	2%	3%
	F*	38	Left	3.16	65	Profound SNHL	SRS	No	No	None	0%	0%
	F*	38	Right	3.28	26	Profound SNHL	SRS	No	No	None	16%	2%

[DOD = duration of deafness, WRS = word recognition score, AZ-Bio = open set speech perception test].
*Represents a single patient with two implanted ears.

Table 2. Cochlear Implant Performance on Patients with NF2 who received observation therapy versus radiosurgery treatment. The "Observation" group is displayed along the top row while the "Radiosurgery" group is displayed along the bottom row.

Group	Study	Participant Demographics: Participant	Gender	Age	Implanted Ear	Tumor Size (cm)	DOD (months)	Post-Implant Outcome Measures: F/U (months)	PTA	WRS in Quiet	CNC - Phonemes	CNC -Words	CUNY - Quiet	HINT
OBSERVATION GROUP	Mukherjee et al., 2013	1	F	79	Left	1.5	26	-	-	-	-	-	93%	-
		2	F	61	Right	0.13	6	-	30	-	-	-	-	-
	Lustig et al., 2006	3	M	35	NR	-	6	28	55	0%	-	-	0%	-
		4	M	51	NR	-	6	40	35	-	6%	-	-	-
		5	M	16	NR	-	18	30	40	0%	-	-	0%	-
	Tran Ba Huy, 2009	6	F	51	Right	-	84	6	-	68%	-	-	-	81%
		7	F	17	Left	-	-	6	-	87%	-	-	-	98%
	Carlson et al., 2012	8	F	-	NR	-	-	-	18	-	51%	28%	66%	63%
RADIOSURGERY GROUP	Mukherjee et al., 2013	1	M	44	Left	1.5	-	-	-	-	-	-	0%	-
		2	F	72	Left	-	24	-	-	-	-	-	0%	-
		3	F	20	Left	3.7	84	-	-	-	-	-	0%	-
	Lustig et al., 2006	4	F	41	NR	-	36	17	55	46%	-	-	-	-
		5	M	50	NR	-	24	18	35	46%	-	-	-	98%
	Tran Ba Huy, 2009	6	M	26	Right	-	-	12	-	96%	100%	-	-	-
	Carlson et al., 2012	7	M	-	NR	-	-	-	18	-	67%	46%	100%	95%
		8	F	-	NR	-	-	-	20	-	93%	86%	-	-
		9	F	-	NR	-	-	-	19	-	-	-	-	-
	Current Study	10	M	77	Left	0.10	240	12	22	28%	55%	28%	-	34%
		11*	F	38	Left	3.16	65	12	25	52%	65%	40%	-	45%
		11*	F	38	Right	3.28	26	12	25	8%	37%	24%	-	45%

["-" = No data available, NR = Not Reported, DOD = duration of deafness, PTA = pure tone average, WRS = word recognition score, CNC = consonant-nucleus-consonant, CUNY = City University of New York, HINT = hearing in noise test].
*Represents a single patient with two implanted ears.

cated a flat, profound sensorineural hearing loss in both ears. Aided speech perception scores were 2% for CNC – words and 16% for CNC - phonemes. Aided speech perception score for sentences was 3% in the best aided condition as measured on the Az-Bio test (Table 1).[16]

The patient underwent Gamma Knife SRS to the left tumor with 12.5 Gy to the 50% isodose line. Post SRS the patient experienced transient imbalance. He was subsequently implanted in the left ear in an attempt to maximize speech understanding and communication (Table 1). Post-implant pure tone average was within the normal range of hearing in the implanted ear. Twelve month post-implant speech scores are summarized in Table 2.

Literature Review

A total of 19 individual cases were reported, including the 2 case studies where 3 ears were discussed (Table 2).[3,4,5,6,17] Each study was analyzed to remove duplicate entries.

Tumor Observation with CI

Of the 8 patients, the mean age for the observation group was 42.9 with a median age of 41 years. Average reported tumor size was 0.82cm with a ceiling of 1.5cm. The mean reported duration of deafness since diagnosis was 22 months (1.83 years). Of those reported, 50% of patients were implanted on the right ear and 50% were implanted on the left.

While individual scores varied significantly, the mean post-implantation pure tone average was 35.6dB HL (Figure 1). Average word recognition in quiet was 39%. The average CNC score was 29% for phonemes (n=2) and 28% for words (n=1). Mean open-set speech recognition was 40% on the CUNY and 81% on the HINT (Figure 2).

Stereotactic Radiosurgery followed by CI

Of the 11 patients and 12 ears, the mean age for the SRS group was 45.1 with a median age of 41 years. Average reported tumor size was 2.34cm with a ceiling of 3.7cm. The mean reported duration of deafness since diagnosis was 71 months (5.9 years). Of those reported, 29% of patients were implanted on the right ear and 71% were implanted on the left.

The mean post-implantation pure tone average was 27.8dB HL (Figure 1). Average word recognition in quiet was 63%. The average CNC score was 70% for phonemes and 49% for words (Table 2). Mean open-set speech recognition was 25% on the CUNY and 63% on the HINT (Figure 2).

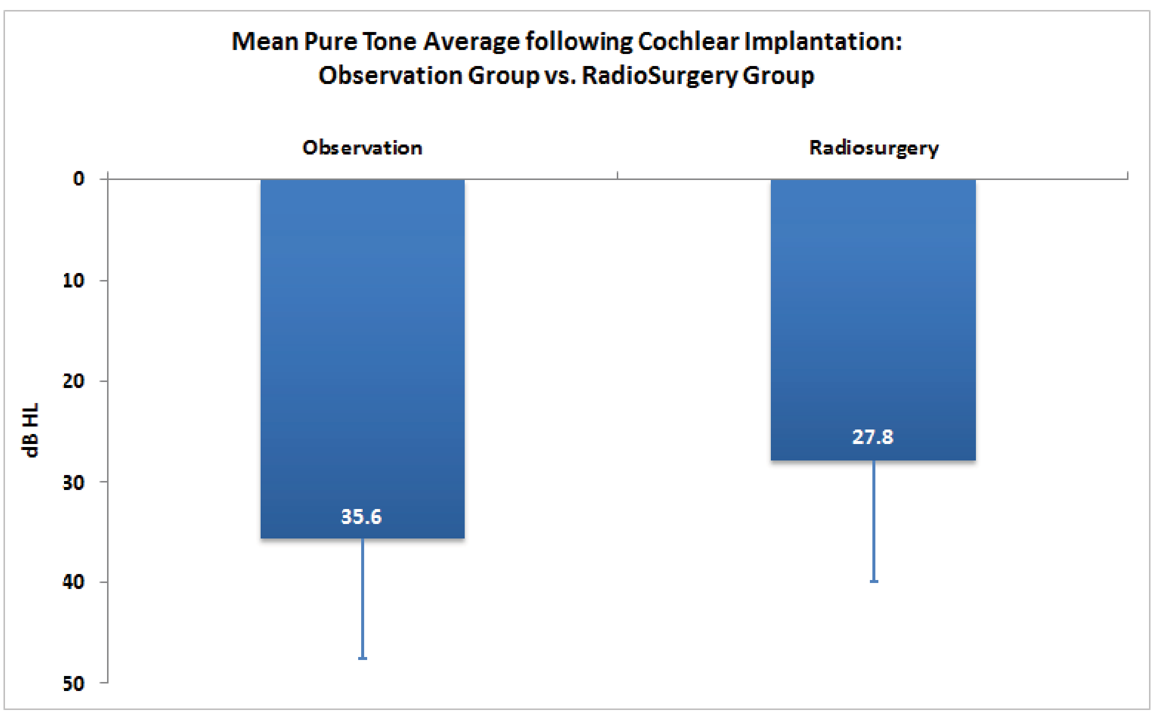

Figure 1. Mean pure tone averages following cochlear implantation by treatment group (Observation versus Stereotactic Radiosurgery) as measured via the implanted ear.

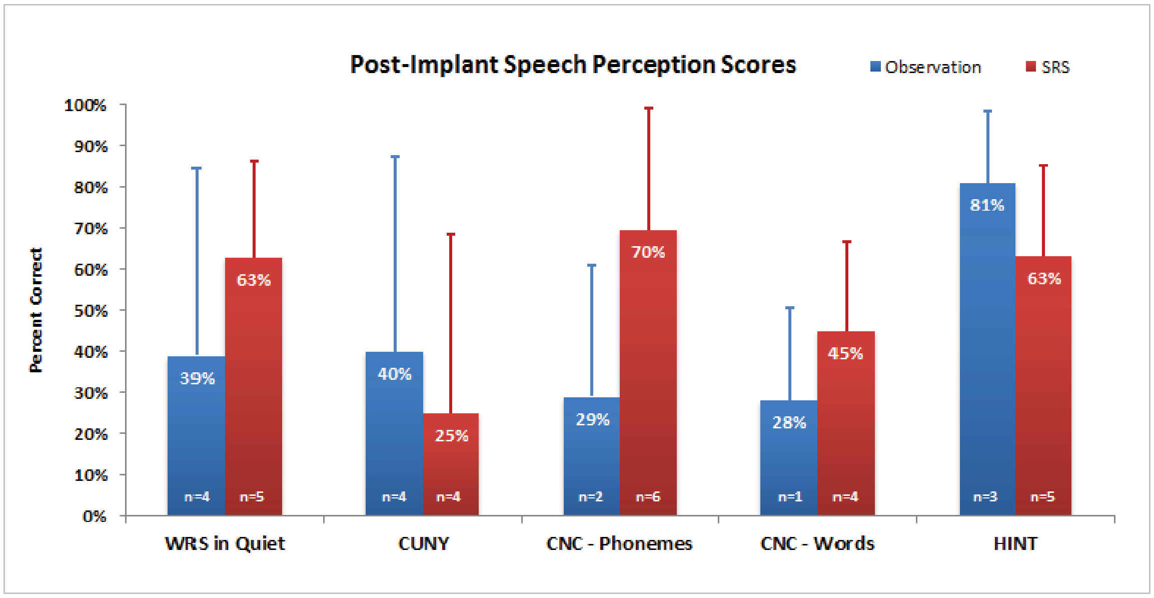

Figure 2. Mean speech perception scores following cochlear implantation by treatment group (Observation versus Stereotactic Radiosurgery).

[DOD = duration of deafness, WRS = word recognition score, CNC = consonant-nucleus-consonant, CUNY = City University of New York, HINT = hearing in noise test].

The post-operative outcomes from both the observation and radiosurgery groups were comparable following cochlear implantation. Both groups achieved similar post-implant pure tone thresholds as measured in sound field through the CI. Additionally, open-set speech perception scores indicated no significant differences between the two cohorts; however, comparable data sets were limited. The largest variance between groups was duration of deafness with the radiosurgery group reporting approximately 3 times the duration of hearing loss compared to the observation group prior to cochlear implantation.

DISCUSSION

As technology progresses, the treatment options for patients with NF2 have grown. Auditory rehabilitation expanded to include ABI and now more commonly CI.[12,17,18,19] While limiting tumor growth is a priority, hearing preservation and communication are also paramount.[11] As a result, surgeons must delicately balance patient safety with quality of life when considering best management.

Although there is a large degree of variability in the limited available data, the outcomes provided in this paper were in line with published measures.[3,4,5,6,17] The findings support cochlear implantation, with significant improvement in open-set speech recognition in both cohorts of patients with NF2; however far less than the 30% to 50% net improvement one might expect from conventional CI recipients.[3,20,21] The available literature, including the findings from this study, demonstrate that radiosurgery followed by cochlear implantation can be a viable treatment of the tumor without negative impact on audition and speech perception as compared to exclusively observed tumors with ipsilateral implantation. However, patients in this cohort should receive extensive pre-surgical counseling on realistic expectations and the potential limiting factors of cochlear implantation relative to NF2.

Implantation raises the important concern regarding surveillance. This is for both vestibular as well as other common NF2 associated tumors. The tissues ipsilateral to the magnet will be obscured for several centimeters (up to 7) on magnetic resonance imaging (MRI) owing to interference with the magnet contained in the scanner.[22,23] This further endorses the impetus for tumor management prior to hearing restorative surgery. Post CI imaging can be facilitated by removing the magnet prior to imaging or with use of a low Tesla MRI.

Risks associated with post CI MRI include; device motion, thermal damage to the soft tissues or cochlea proper, degaussing the internal magnet or other device malfunction.

Long term follow-up and additional research is needed in this area as the instances of patients with NF2 who have a CI are limited. Both tumor and SRS effects may be dynamic and have a late impact on audition. It is hypothesized that SRS tumor control is mediated through endothelial injury as opposed to direct tumor cell damage. [24, 25]

However, the cochlea is known to be radiosensitive with suggestion of both stria vascularis and outer hair cell damage when exposed to radiation exceeding 4Gy.[24, 25]

Others have found spiral ganglion cell loss. Inferring from the above, CI should be a durable form of hearing restoration following SRS as it does not require either the stria vascularis nor the outer hair cells for efficacy. While counter intuitive, this outcome accords with existing literature of early SRS for spontaneous schwannomas with evidence for mitigating future tumor effects on non-CI related, post SRS hearing.[4,5] Work by Neff in 2007 found limited late deterioration in CI function post subtotal tumor resection, however extrapolation to the non-surgical condition is problematic.[26]

SRS in NF2 should be assumed with caution and full disclosure as average control rates of approximately 85% suggest the possibility of additional treatment.[27,28] Although rare, there is known increased risk of malignant transformation in VS treated with radiosurgery.[29] The literature suggests that upwards of half of these transformations occur in patients with NF2, although only accounting for a fraction of all VS.[29] Further, NF2 tumor control rates with SRS are more muted than in sporadic tumors. This does not exclude SRS but stresses the complexity in management.

This study did not include patients with subtotal/near total tumor resection with preservation of the cochlear nerve with either simultaneous or delayed cochlear implantation. Our site has no experience with this treatment. The literature in this field is expanding following work by Cueva and Hoffman in the early 1990s illustrating intact electrical promontory stimulation following microsurgical resection.[30, 31] However, microsurgical tumor resection often results in mechanical or thermal injury to the cochlear nerve or labyrinthine artery. The sensory nerve is far more fallible than the motor facial nerve. Hence routine cochlear nerve preservation following surgical resection is challenged.

Study limitations are framed by retrospective and incomplete data acquisition across previous studies. The duration of deafness varied greatly among subjects and disparate speech perception tests were employed. There is limited follow up data and of those reporting, the duration is nominal. This highlights the need that all future published papers include an expansive open-set speech perception test battery at discrete testing intervals in order to achieve a better comparison between patients. Consideration should be made for the expansion of the typical speech perception test battery to create consistency across studies and ensure a valid assessment of the auditory system. This would offer surgeons more information when making critical decisions to ensure the longevity and quality of life for patients with NF2.

CONCLUSION

This study accords with current literature finding that patients with NF2 can achieve open-set speech recognition after undergoing SRS for tumor control with sequential CI. Further, no significant differences in speech perception scores were found between cochlear implant patients undergoing observation therapy versus SRS. This suggests that upfront tumor control does not impair CI function and further minimizes the considerations around tumor surveillance and complicated post implantation tumor management.

List of Abbreviations

NF2: Neurofibromatosis Type II
SRS: Stereotactic Radiosurgery
CI: Cochlear Implant
ABI: Auditory Brainstem Implant
CNC: Consonant-Nucleus-Consonant
CUNY: City University of New York
HINT: Hearing In Noise Test
MR: Magnetic Resonance
MRI: Magnetic Resonance Imaging
dB HL: Decibel Hearing Level

DECLARATIONS

Ethics approval and consent to participate:

University of Manitoba Research Ethics Board (REB) Registry Number: H2015:209 (HS18623), Registered May 25, 2015.

Consent for publication

Participants signed informed consent prior to surgery and data collection.

Availability of data and material:

Please contact author for data requests.

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Authors' disclosure of potential conflicts of interest
Jordan B. Hochman is a Consultant for Advanced Bionics. Other authors have nothing to disclose.

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Conception and design: Justyn Pisa and Jordan Hochman
Data collection: Jordan Hochman, Jacob Sulkers, Michael West, and James Butler
Data analysis and interpretation: Justyn Pisa
Manuscript writing: Justyn Pisa and Jordan Hochman
Final approval of manuscript: Justyn Pisa, Jordan Hochman, Jacob Sulkers, Michael West, and James Butler

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