

National Prion Disease Pathology Surveillance Center

2026 Update & Panel



SCHOOL OF MEDICINE

CASE WESTERN RESERVE
UNIVERSITY

Summary

- Provide a brief overview of the NPDPS
- Provide some basic info on our findings
- Q&A Panel with NPDPS faculty and staff



SCHOOL OF MEDICINE

CASE WESTERN RESERVE
UNIVERSITY

NPDPS Mission

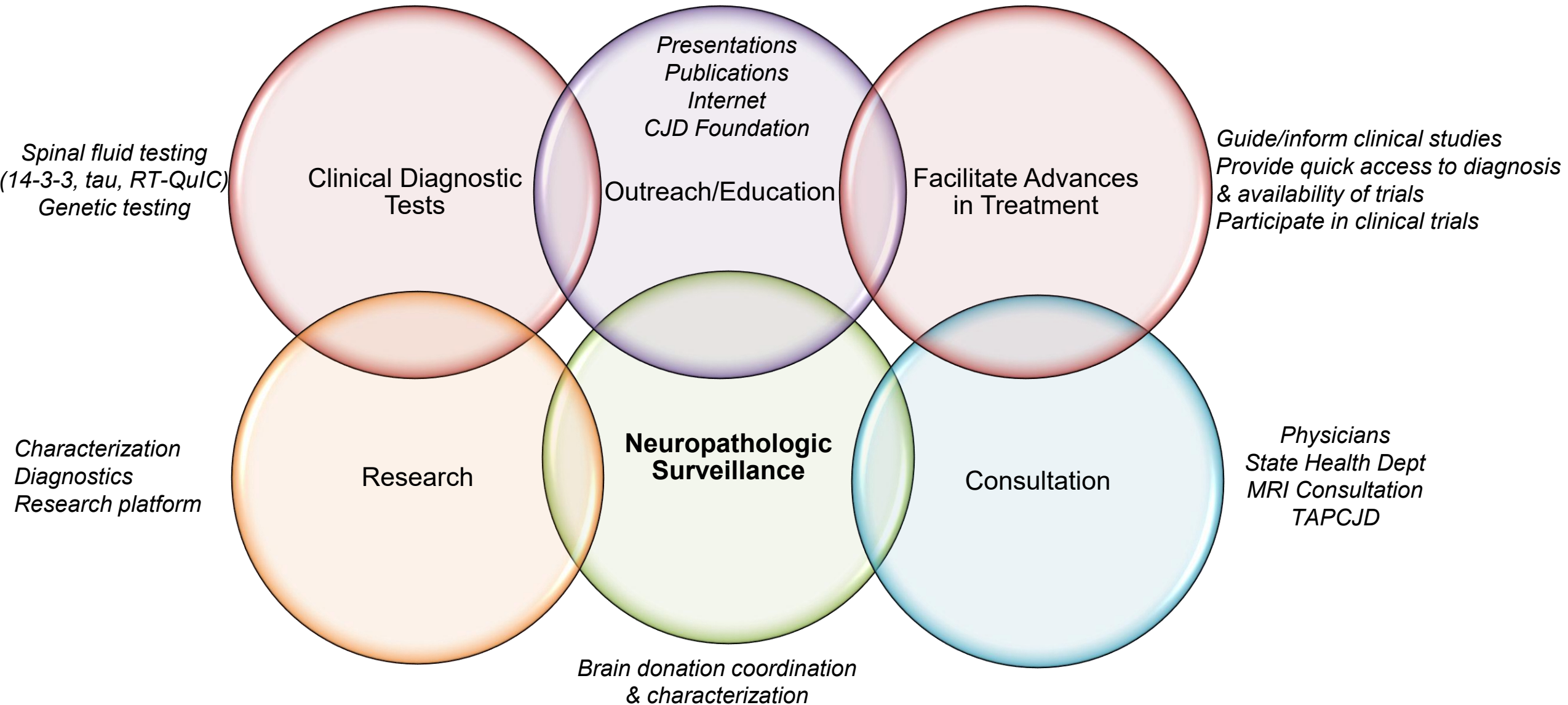
- Started in 1997 at Case Western Reserve University in response to variant CJD by CDC in collaboration with Dr. Pierluigi Gambetti
- Neuropathologic surveillance activities are funded by CDC
 - Allows for a definitive diagnosis of prion disease
 - Determines the type of prion disease
 - Allows for neuropathologic and biochemical surveillance for novel forms of the disease
- Important focus is on preventable & potentially novel forms of prion disease (e.g., variant CJD, iatrogenic CJD, and novel acquired prion diseases)



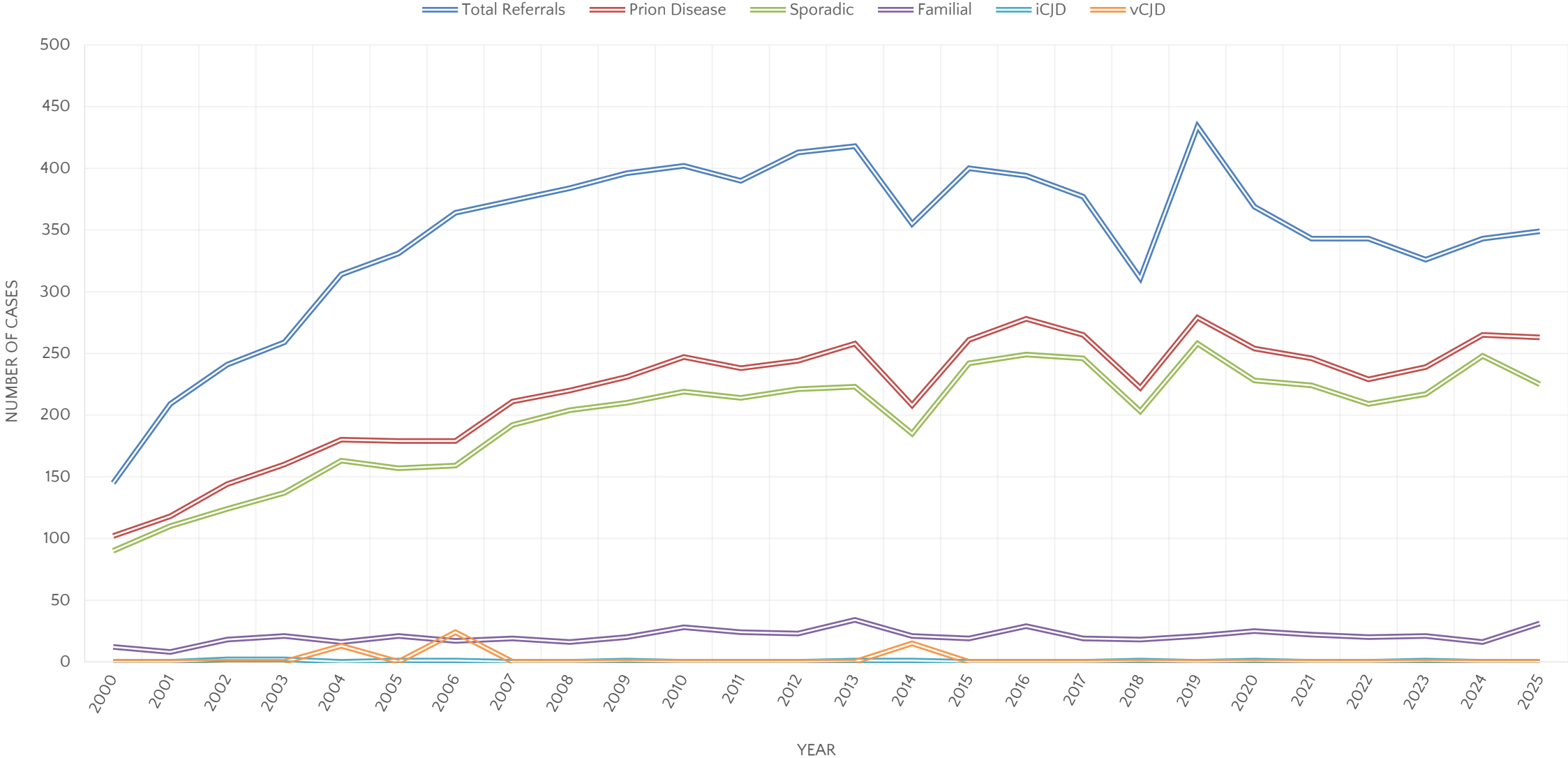
SCHOOL OF MEDICINE

CASE WESTERN RESERVE
UNIVERSITY

Aspects of the NPDPS

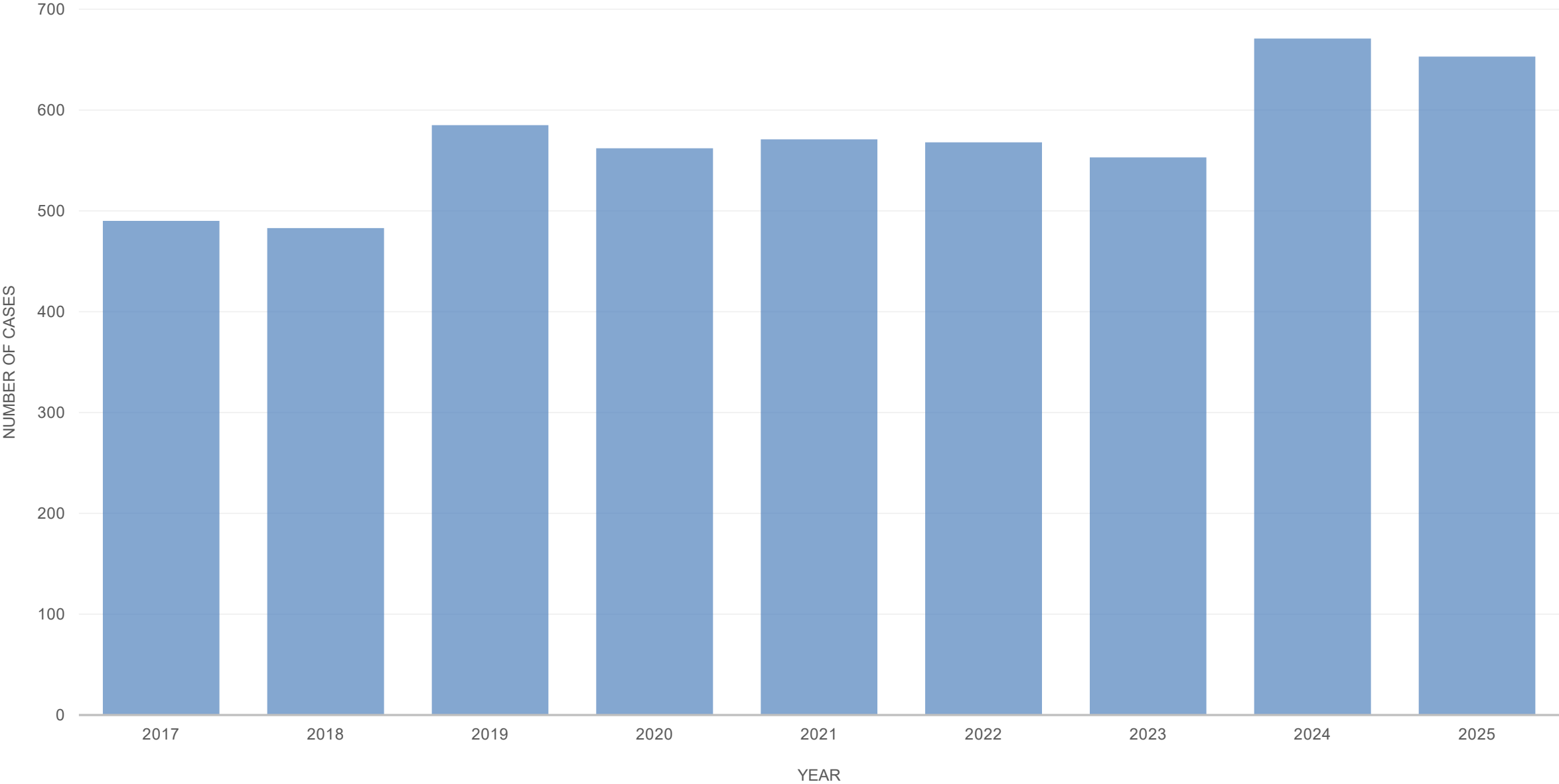


NEUROPATHOLOGIC ANALYSES AT NATIONAL PRION DISEASE
PATHOLOGY SURVEILLANCE CENTER
NUMBER OF CASES PER YEAR



vCJD: 4 cases (all exposed outside the US)
iCJD: 15 cases, mostly from c-hGH

Probable & Definite Prion Disease Cases By Year



Definite = Autopsy confirmed
Probable = Autopsy confirmed + Positive RT-QuIC cases that did not go to autopsy

Sporadic Prion Disease Distribution in the US

Sporadic Subtype	Percentage, 2025
MM1	39.5
MM1-2	11.6
MM2	3.7
MV1	6.1
MV1-2	11.6
MV2	9.8
sFI	0.5
VPSPr	1.9
VV1	2.3
VV1-2	4.7
VV2	8.4



SCHOOL OF MEDICINE

CASE WESTERN RESERVE
UNIVERSITY

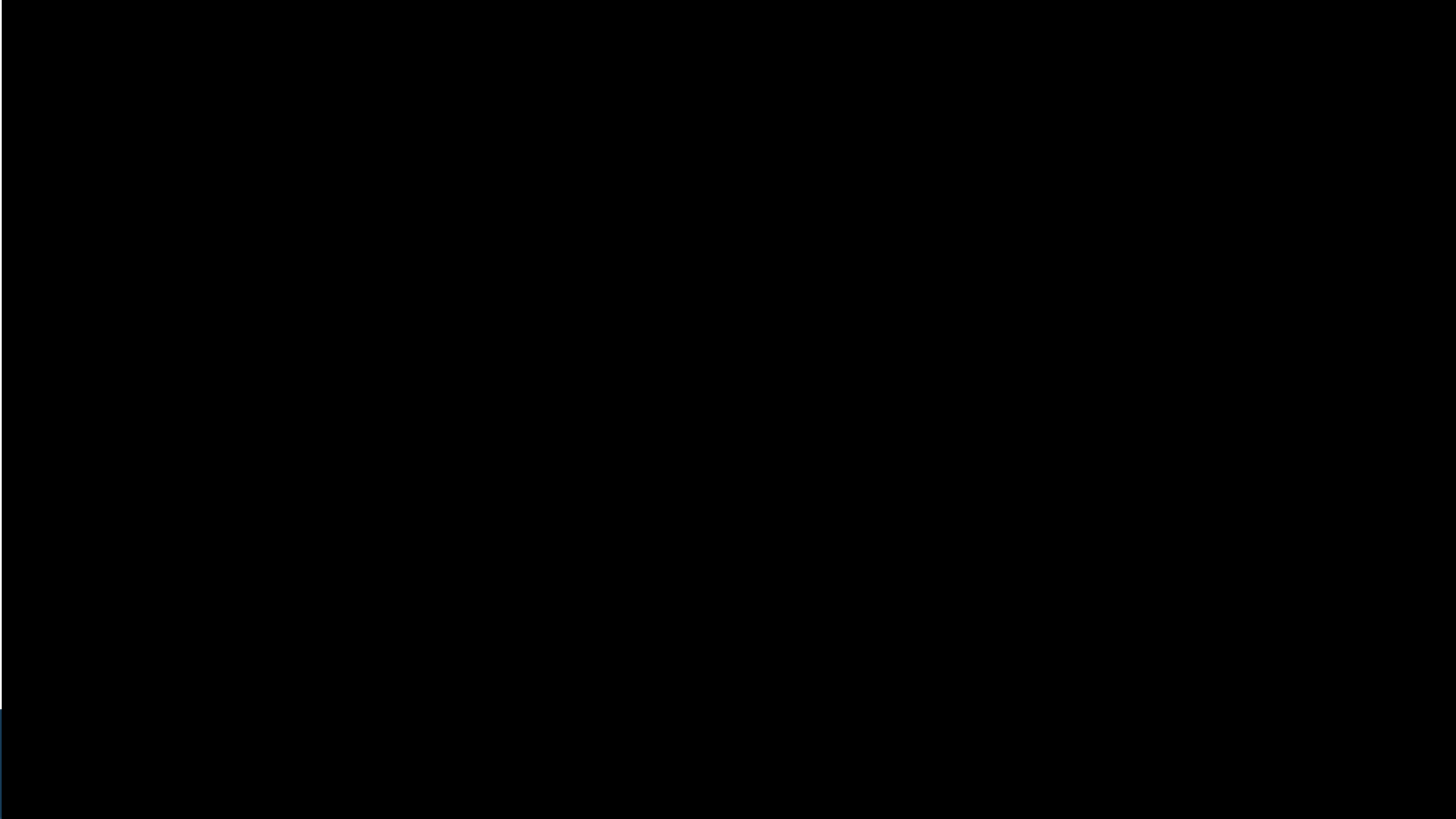
Genetic Prion Disease Mutations in the US

Mutation	Total	Mutation	Total
E200K-129M	346	G114V-129M	3
Insertions (OPRI)	104	R208H-129V	3
V210I-129M	92	V180I-129V	3
D178N-129M	76	E196K-129M	2
P102L-129M	64	H187R-129V	2
E200K-129V	59	P102L-129V	2
D178N-129V	38	Q217R-129V	2
T188R-129V	31	A224V-129V	1
A117V-129V	25	G131E-129M	1
F198S-129V	11	G94S-129V	1
H187R-129M	10	H187A-129M	1
V180I-129M	10	N171S-129V	1
P105S-129V	6	Q160X-129M	1
R208H-129M	6	Q186X-129V	1
G54S-129M	5	R148H-129V	1
T183A-129M	5	S245P-129M	1
R148H-129M	4	T183A-129V	1
V203I-129M	4	T188A-129V	1
V203I-129V	4	T188K-129V	1
A133V-129M	3	T193I-129M	1
E196A-129M	3	V189I-129M	1
E200G-129V	3	P105S*	1
		Deletion (Total)	1
Subdivision of Insertions			
Mutation	Total	Mutation	Total
5 OPRI-129V	24	9 OPRI-129V	5
6 OPRI-129M	22	1 OPRI-129M	3
5 OPRI-129M	15	9 OPRI-129M	1
4 OPRI-129V	11	1 OPRI-129V	1
6 OPRI-129V	10	8 OPRI-129M	1
2 OPRI-129M	10	8 OPRI*	1
Subdivision of Deletions			
2 Deletions-129M	1		
Cases could not be haplotyped (P105S, 8 OPRI)			



SCHOOL OF MEDICINE

CASE WESTERN RESERVE
UNIVERSITY





cjd

FOUNDATION

NPDPSC Website

cjdsurveillance.com

Content

- Information on offered services
- Information on prion disease
- Information on enrolling clinical trials
- Quarterly updates to surveillance data
- Articles & talks from the NPDPSC with lay summaries (Newsfeed)

Chat with our publications

Clinical Testing

The NPDPSC offers various laboratory services for patients suspected of having a prion disease.



Diagnostic Activities

Laboratory services of the center, including our lab testing directory, test requisition forms, resources for healthcare professionals, and more.

 Treatment Trials

 Newsfeed

NPDPSC INTERACTIVE
PUBLICATIONS

Use our interactive publications chat, powered through NotebookLM, to explore NPDPSC research, published findings, and prion disease topics using natural language questions. Users can quickly identify relevant studies, mutations, diagnostic methods, and other research topics across the NPDPSC publication library.

Chat With NPDPSC
Publications Here



SCHOOL OF MEDICINE

CASE WESTERN RESERVE
UNIVERSITY

Huge Thank You!

- CJD Foundation
- Akin
- Families
- Friends (fellow scientists/collaborators)



SCHOOL OF MEDICINE

CASE WESTERN RESERVE
UNIVERSITY

NPDPS Panel

- Eriton Cunha, PhD: Western blot lab
- Gabriella DelPrete: Tissue coordinator
- Danielle Jordan: Brain donation program manager
- Keisi Kotobelli, MS, GStat: Data manager
- Liam Wissinger: Digital pathology manager
- You!



SCHOOL OF MEDICINE

CASE WESTERN RESERVE
UNIVERSITY