Table S1. STARD Checklist for the Reporting of Studies of Diagnostic Accuracy.

Section & Topic	No	Item	Reported on page #
TITLE OR ABSTRACT			
	1	Identification as a study of diagnostic accuracy using at least one measure of accuracy (such as sensitivity, specificity, predictive values, or AUC)	1
ABSTRACT			
	2	Structured summary of study design, methods, results, and conclusions (for specific guidance, see STARD for Abstracts)	3
INTRODUCTION			
	3	Scientific and clinical background, including the intended use and clinical role of the index test	4
	4	Study objectives and hypotheses	4
METHODS			
Study design	**5**	Whether data collection was planned before the index test and reference standard were performed (prospective study) or after (retrospective study)	5
Participants	**6**	Eligibility criteria	5
	7	On what basis potentially eligible participants were identified (such as symptoms, results from previous tests, inclusion in registry)	5
	8	Where and when potentially eligible participants were identified (setting, location and dates)	5
	9	Whether participants formed a consecutive, random or convenience series	5
Test methods	**10a**	Index test, in sufficient detail to allow replication	5
	10b	Reference standard, in sufficient detail to allow replication	6
	11	Rationale for choosing the reference standard (if alternatives exist)	n/a
	12a	Definition of and rationale for test positivity cut-offs or result categories of the index test, distinguishing pre-specified from exploratory	8
	12b	Definition of and rationale for test positivity cut-offs or result categories of the reference standard, distinguishing pre-specified from exploratory	8
	13a	Whether clinical information and reference standard results were available to the performers/readers of the index test	5
	13b	Whether clinical information and index test results were available to the assessors of the reference standard	5
Analysis	**14**	Methods for estimating or comparing measures of diagnostic accuracy	6
	15	How indeterminate index test or reference standard results were handled	n/a
	16	How missing data on the index test and reference standard were handled	n/a
	17	Any analyses of variability in diagnostic accuracy, distinguishing pre-specified from exploratory	n/a
	18	Intended sample size and how it was determined	6
RESULTS			
Participants	**19**	Flow of participants, using a diagram	Figure S1
	20	Baseline demographic and clinical characteristics of participants	Table 1
	21a	Distribution of severity of disease in those with the target condition	Table 1
	21b	Distribution of alternative diagnoses in those without the target condition	n/a
	22	Time interval and any clinical interventions between index test and reference standard	n/a
Test results	**23**	Cross tabulation of the index test results (or their distribution) by the results of the reference standard	Figure S2 Figure S3
	24	Estimates of diagnostic accuracy and their precision (such as 95% confidence intervals)	7
	25	Any adverse events from performing the index test or the reference standard	n/a
DISCUSSION			
	26	Study limitations, including sources of potential bias, statistical uncertainty, and generalisability	11
	27	Implications for practice, including the intended use and clinical role of the index test	11
OTHER INFORMATION			
	28	Registration number and name of registry	12
	29	Where the full study protocol can be accessed	n/a
	30	Sources of funding and other support; role of funders	12

Figure S1. Diagram of the participants' study flow. mtDNA+ and mtDNA- correspond to mtDNA copy numbers above and below the calculated cut-off, respectively.

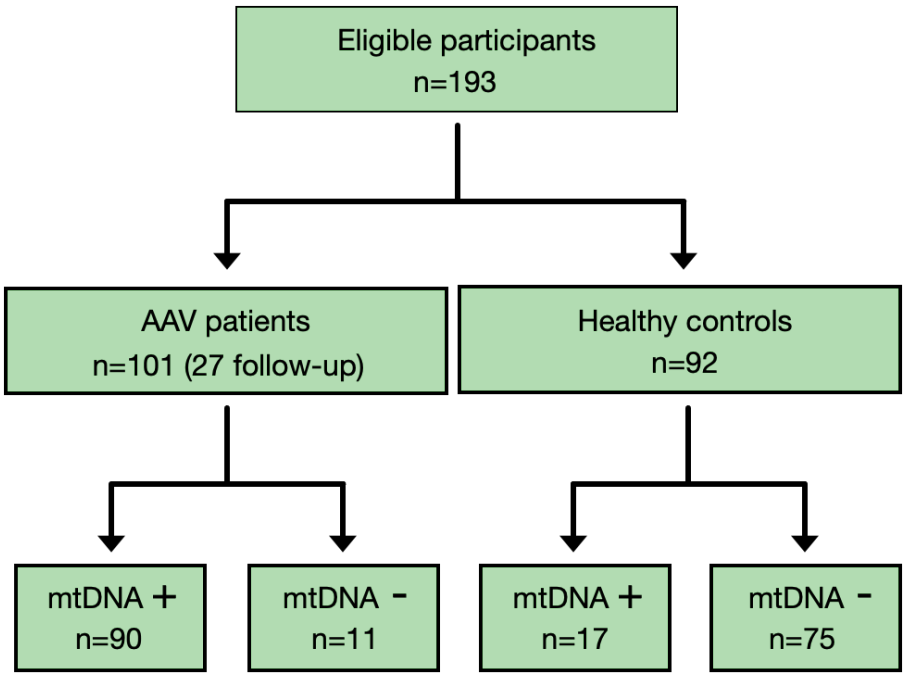

Figure S2. Confusion matrix depicting the performance of mtDNA copy number determination as a biomarker for reliable diagnostic assessment of patients with AAV. Values of sensitivity, specificity, accuracy, precision and negative predictive value (NPV) are also included.

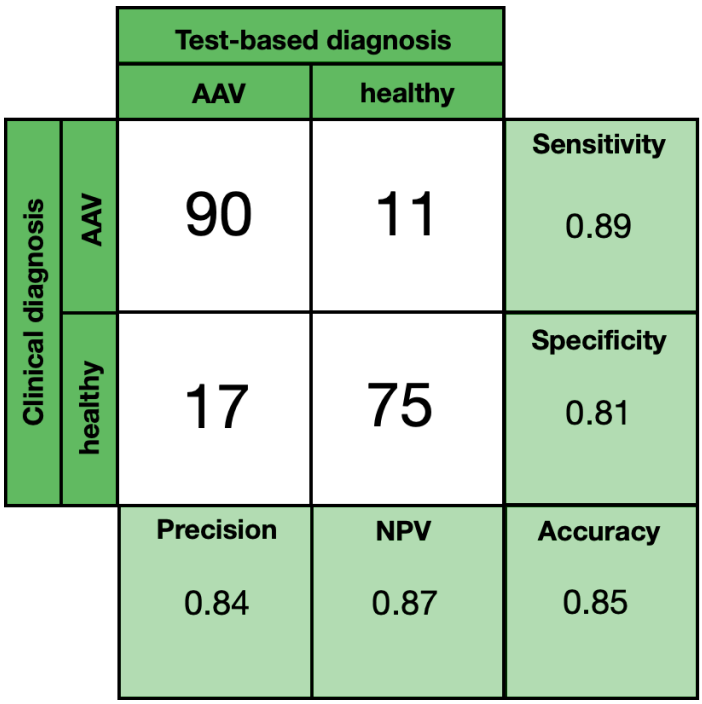

Figure S3. Confusion matrix depicting the performance of mtDNA copy number determination as a biomarker for reliable diagnostic assessment of disease active AAV patients. Values of sensitivity, specificity, accuracy, precision and negative predictive value (NPV) are also included.