

SUPPLEMENTARY MATERIAL FOR

Development of a kit for urine collection on filter paper as an alternative for Pompe disease screening and monitoring by LC-HRMS

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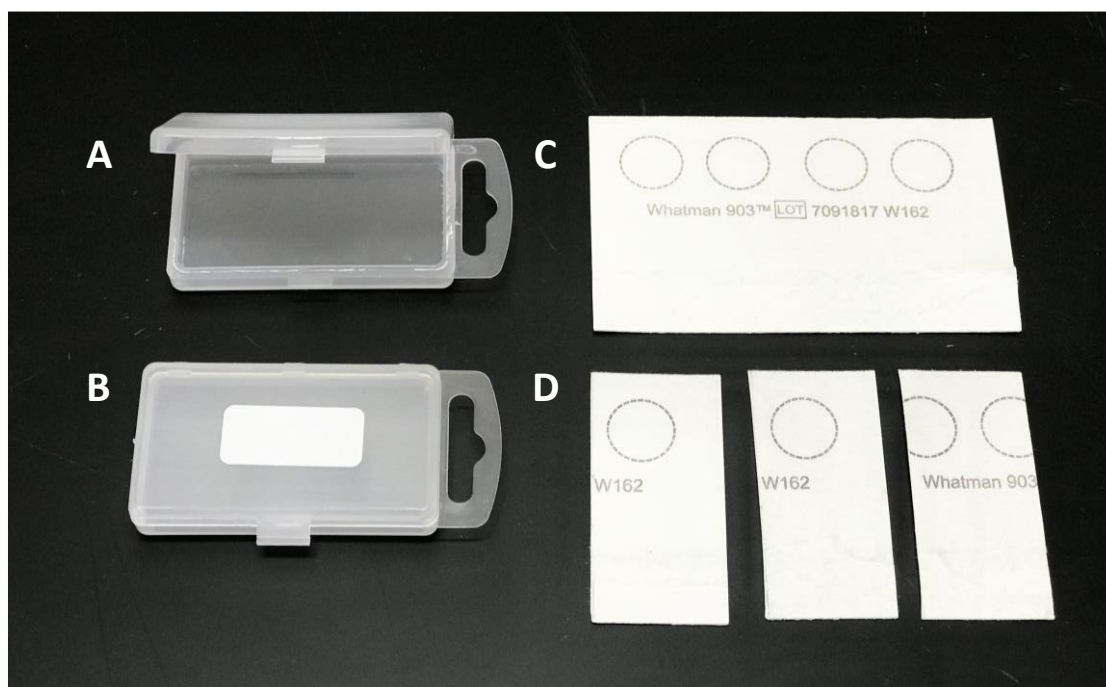


Fig. S1 Front view of the plastic box (width 2.5 cm x length 5.0 cm) (A) open and (B) closed, used in this study for the purpose of collecting, storing and extracting Glc₄ and creatinine using DUFP. The plastic box contains a hinge on the box lid and a safety lock. (C) Filter paper (Whatman 903) commercially available in width 5.0 cm x length 9.0 cm and cut off into three equal parts (width 2.5 cm x length 5.0 cm).

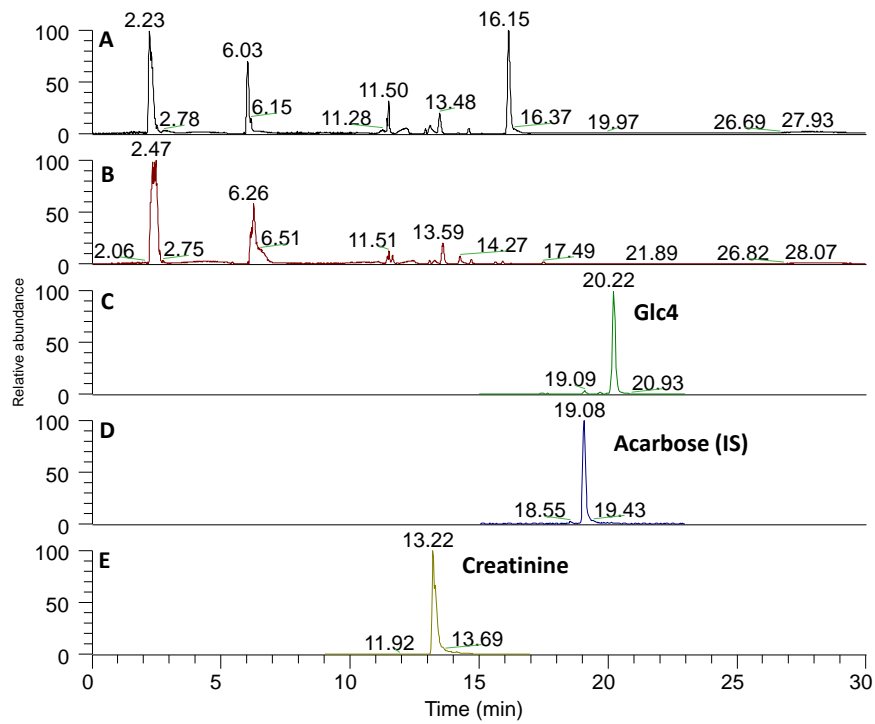


Fig. S2 Representative base peak chromatograms of urine samples from Pompe patients (A) and control group (B) collected on filter paper and typical PRM chromatograms for the (C) Glc4, (D) acarbose IS and (E) creatinine obtained by LC-(ESI)-HRMS.

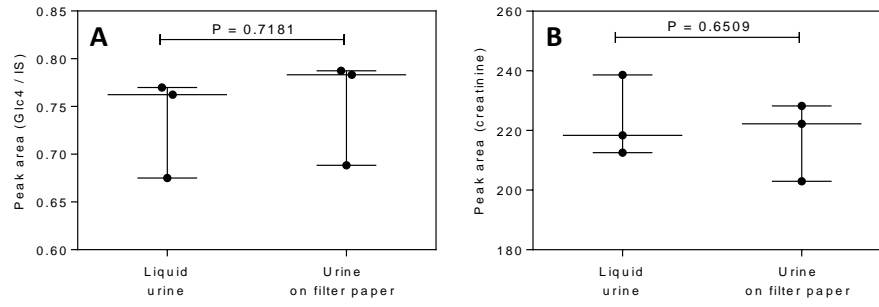


Fig. S3 Comparative boxplot showing no statistically significant difference between (A) Glc4 and (B) creatinine in urine liquid (control) and urine on filter paper mailed to laboratory.

Table S1 Chemical composition of surrogate synthetic urine.

| Chemical composition | Concentration |
|-----------------------------|----------------------|
| Uric acid | 46 mg/L |
| Calcium | 45 mg/L |
| Chloride | 50 mEq/L |
| Creatinine | 237 mg/L |
| Phosphor | 247 mg/L |
| Glucose | 124 mg/L |
| Magnesium | 32 mg/L |
| Potassium | 7 mEq/L |
| Total proteins | 135 mg/L |
| Sodium | 60 mEq/L |
| Urea | 1744 mg/L |