

Polyglucosan body structure in Lafora disease

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Supplementary Information

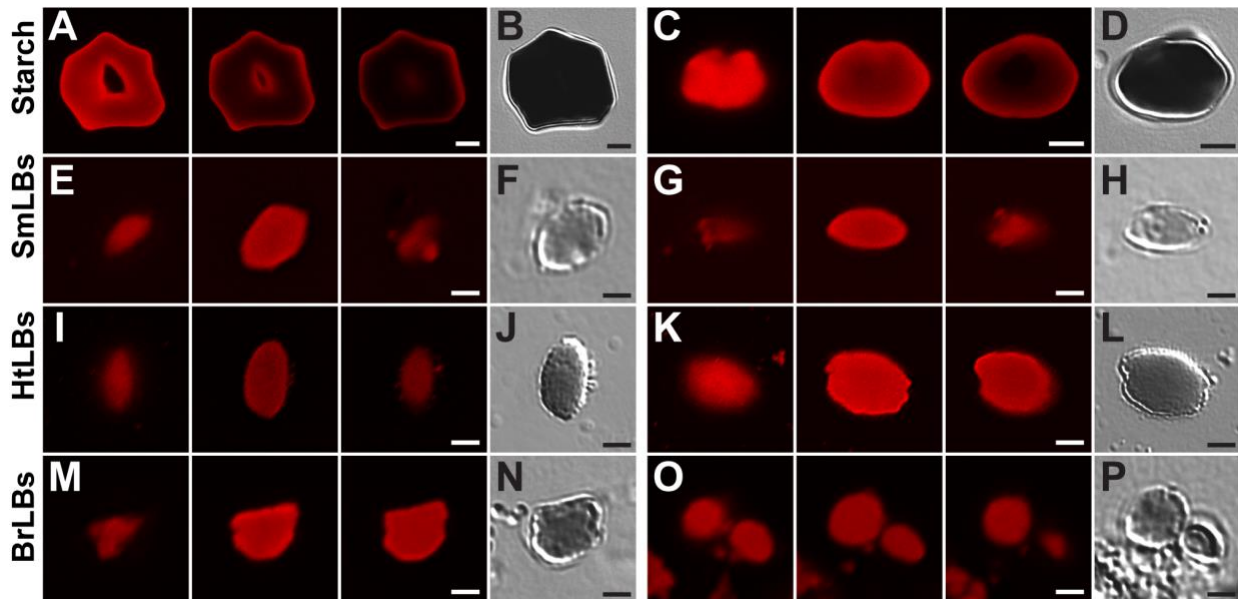


Figure S1. Additional confocal images of starch and LBs. Z-stack 561 nm confocal images of iodine-stained starch (A, C), SmLBs (E, G), HtLBs (I, K), and BrLBs (M, O). Corresponding DIC images for starch (B, D), SmLBs (F, H), HtLBs (J, L) and BrLBs (N, P) are shown. In (A) through (D), scale bars = 5 μm and TRITC panels represent 2.25 μm steps. In (E) through (P), scale bars = 2 μm and panels represent 0.9 μm steps.

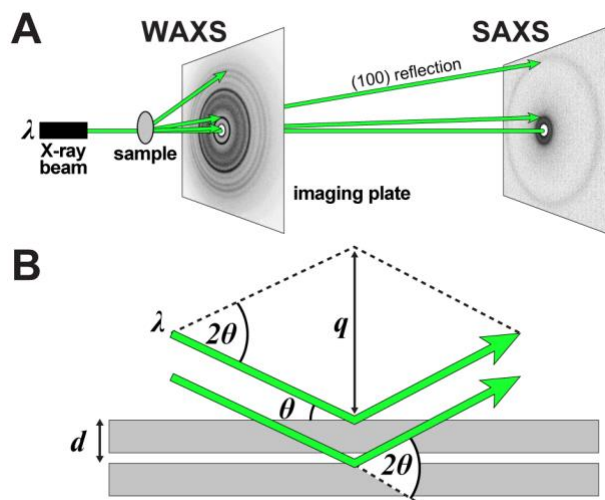


Figure S2. Schematic diagram of SAXS and WAXS experimental setup and theory. (A) X-ray radiation at a specific wavelength (λ) is scattered upon crossing the sample. The scattered X-rays are collected on an imaging plate, producing a diffraction pattern containing concentric rings if the specimen is an isotropic powder. In SAXS, the imaging plate is placed further from the sample to collect the scattering signal at small angles. In WAXS, the imaging plate is closer to the sample to collect the scattering at wide angles. (B) A schematic depiction of Bragg's law. The crystalline units (grey bars, representing either lamellar layers or individual helices) within the sample are separated by a distance, d . Upon interaction with the crystalline material, the incident radiation (with wavelength, λ) is diffracted at an angle (2θ) related to the scattering vector (q) and repeating distance (d) between the crystalline units as described in Equation (1).

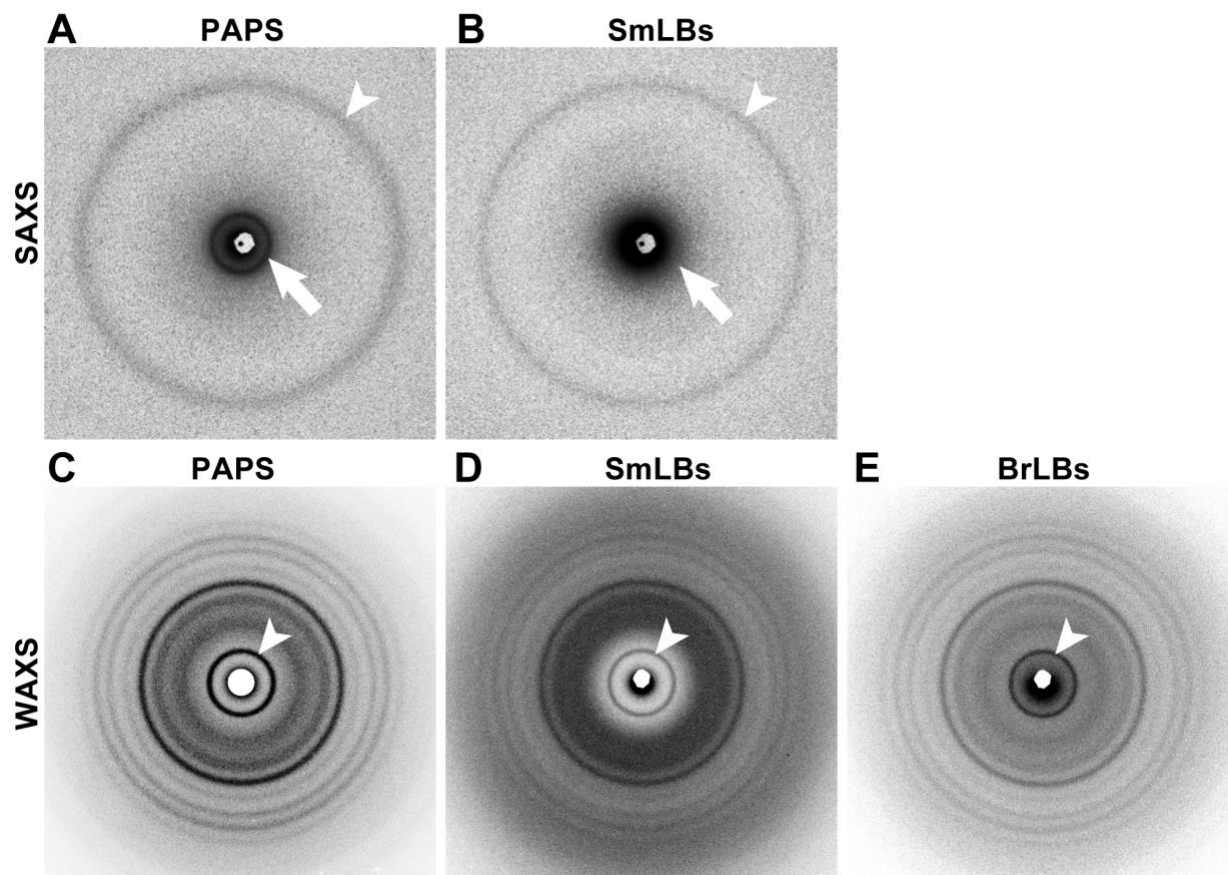


Figure S3. 2D SAXS and WAXS diffraction patterns. (A, B) 2D SAXS diffraction patterns of PAPS (A) and SmLBs (B). The numerical data are plotted in Figure 2B. (C, D, E) 2D WAXS diffraction patterns of PAPS (C), SmLBs (D) and BrLB (E) illustrate a B-type ring distribution, with data plotted in Figure 2C. The white arrow indicate the location of the lamellar reflection at $q = 0.76 \text{ nm}^{-1}$. The arrowheads indicate equivalent reflections at $q = 4.1 \text{ nm}^{-1}$ (A, B) and 5.8° (C, D, and E), which correspond to the same B-type reflection.

SmLBs
30 min 95°C + 3 days 4°C

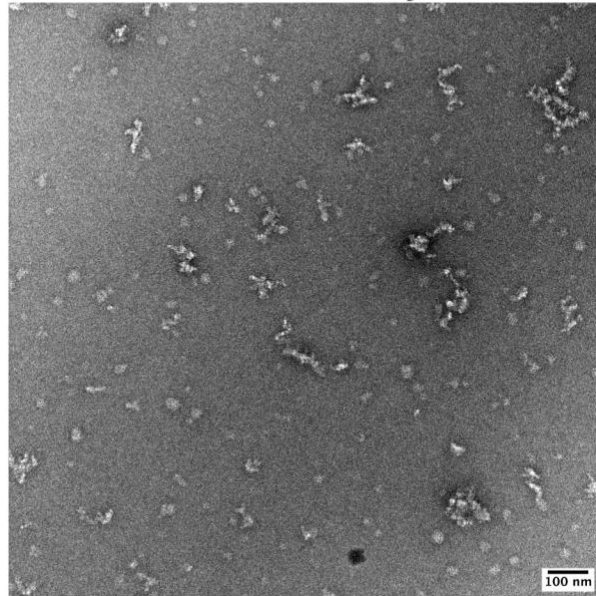


Figure S4. Additional TEM image of heated SmLBs. SmLBs visualized after heating for 30 min at 95 °C and then a 3-day retrogradation period at 4 °C.