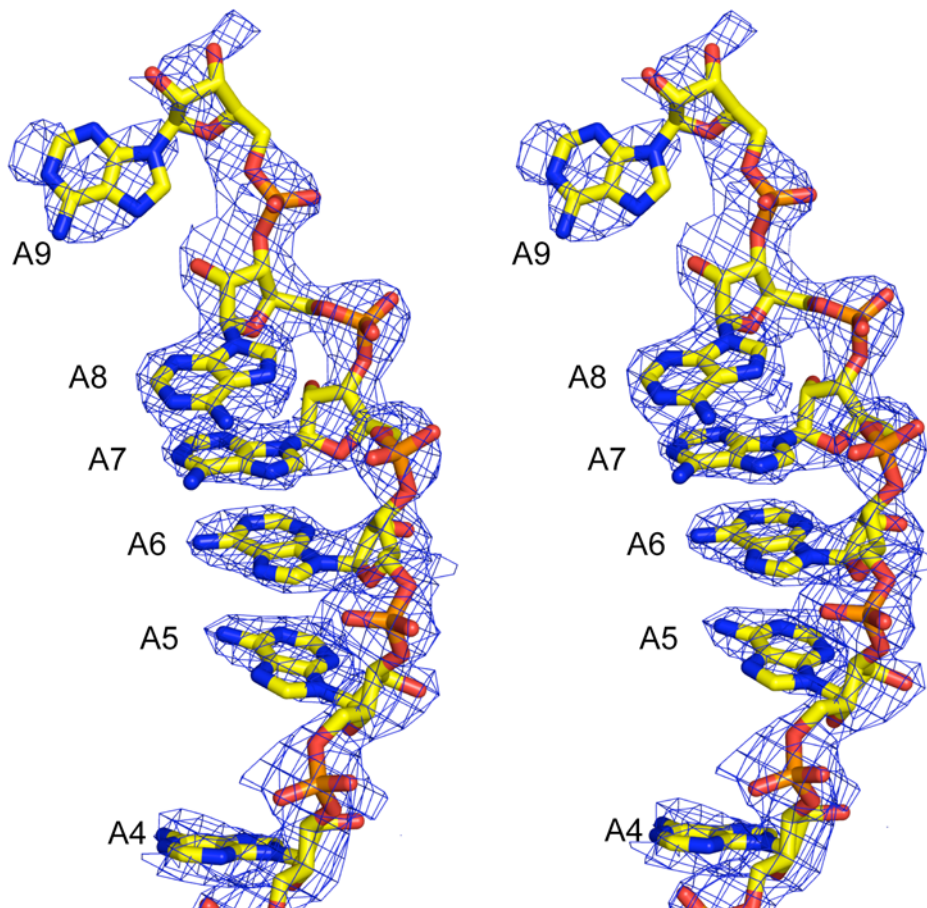**SUPPORTING INFORMATION**

Supplementary Figure 1. The electron density map of the bound ssRNA. Stereo diagram of 2.85Å simulated annealing omit map (blue) contoured at 1.0 σ showing the bound poly(A) of molecule A in the hIghmbp2hd-RNA structure. For simplicity, only the last five nucleotides of the 9-mer ssRNA are shown in stick models.

Supplementary Figure 2. Sequence alignment of helicase domains of Upf1 and Ighmbp2. The amino acids sequences of human (*H.s.*), mouse (*M.m.*) and hamster (*M.a.*) Ighmbp2 were aligned with those of human and Saccharomyces cerevisiae (*S.c.*) Upf1. Residues of human Ighmbp2 associated with disease-causing missense mutations in SMARD1 are marked with "•".

Supplementary Figure 1

Supplementary Figure 2