

Supporting Information

Synthesis of 1,5-Dideoxy-1,5-Iminoribitol-*C*-Glycosides Through a Nitron-Olefin Cycloaddition Domino Strategy: Identification of Pharmacological Chaperones of Mutant Human Lysosomal β -Galactosidase

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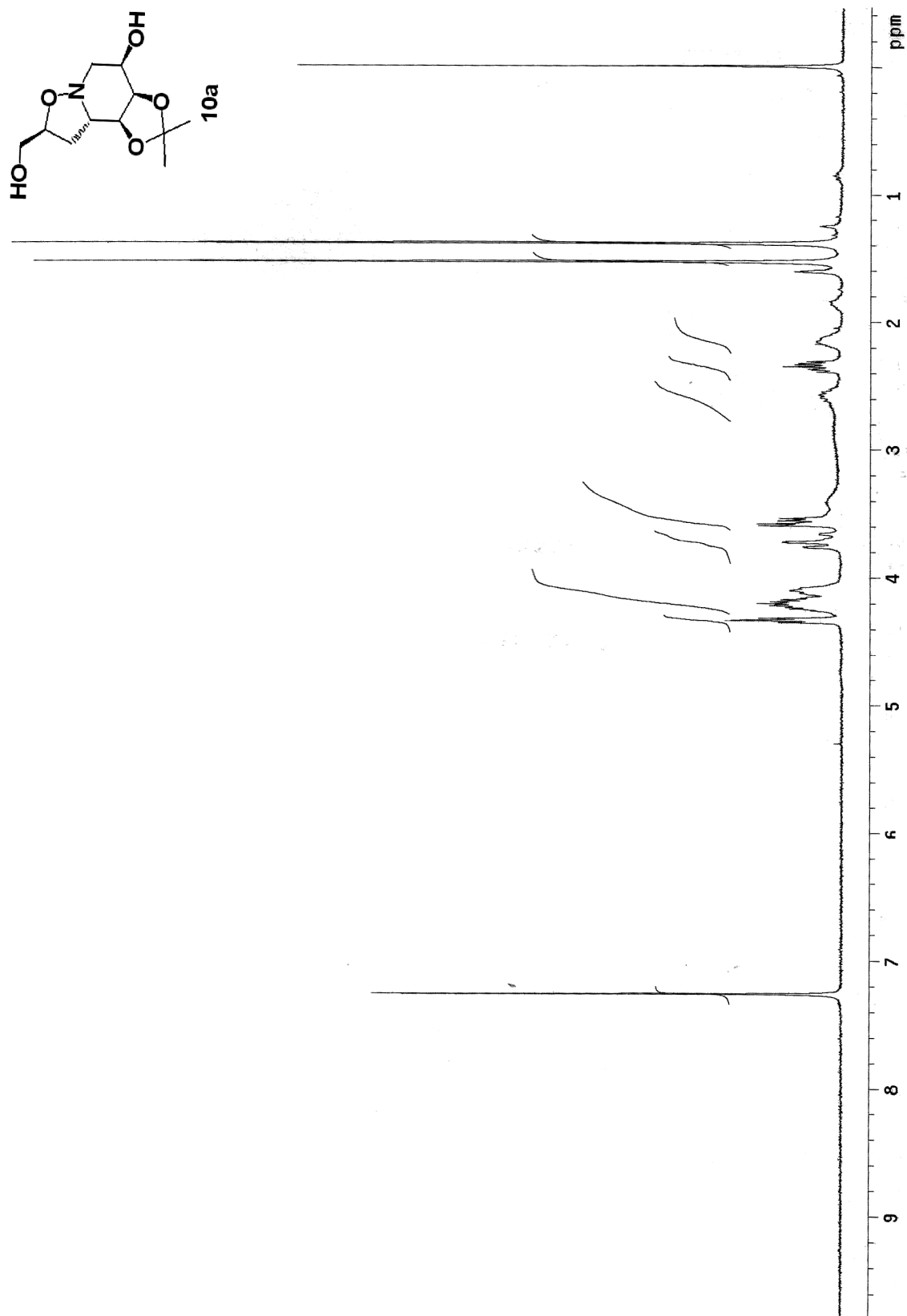
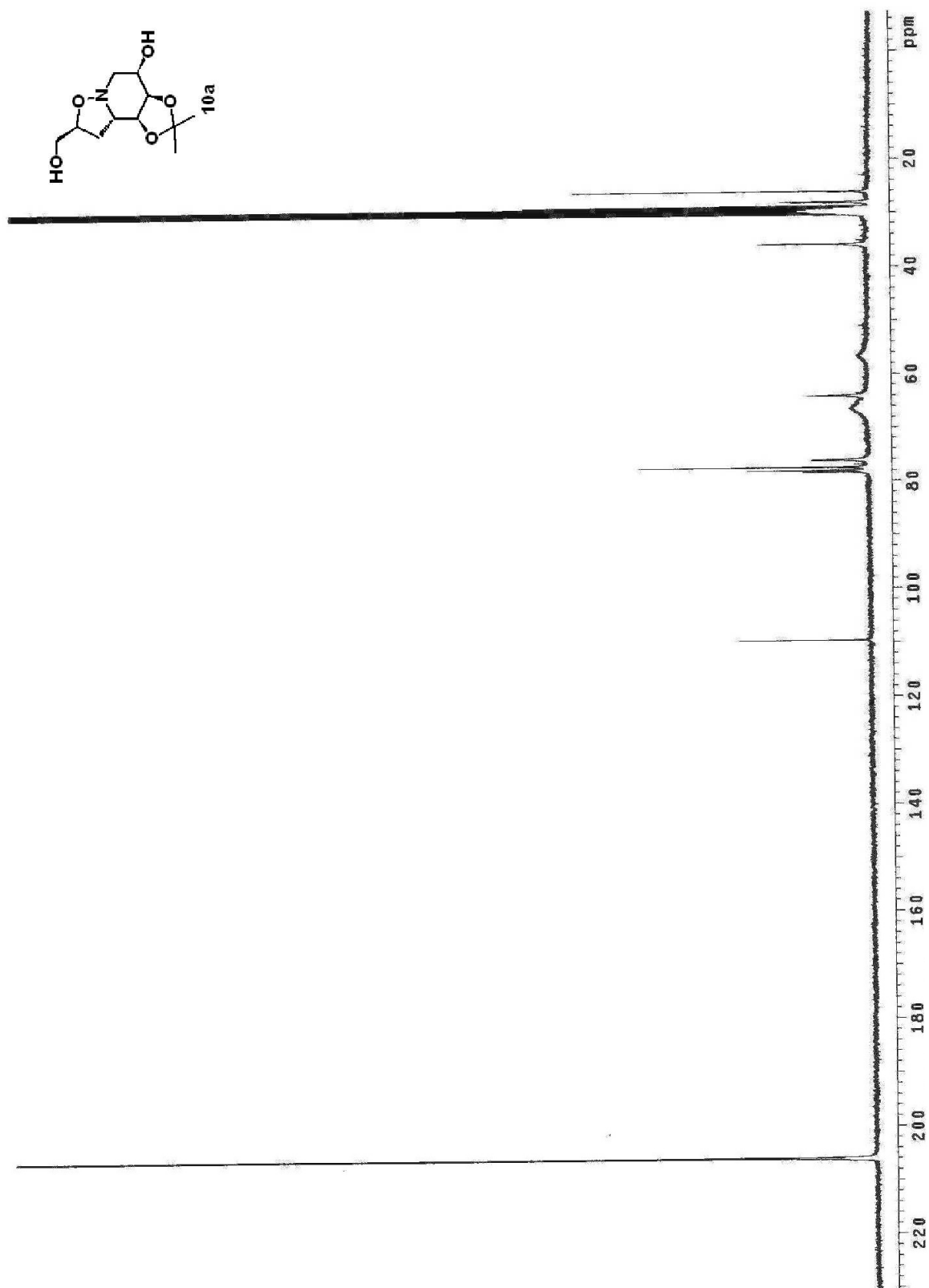


Figure 1S: ^1H (300 MHz, CDCl_3) Spectrum of compound 10a



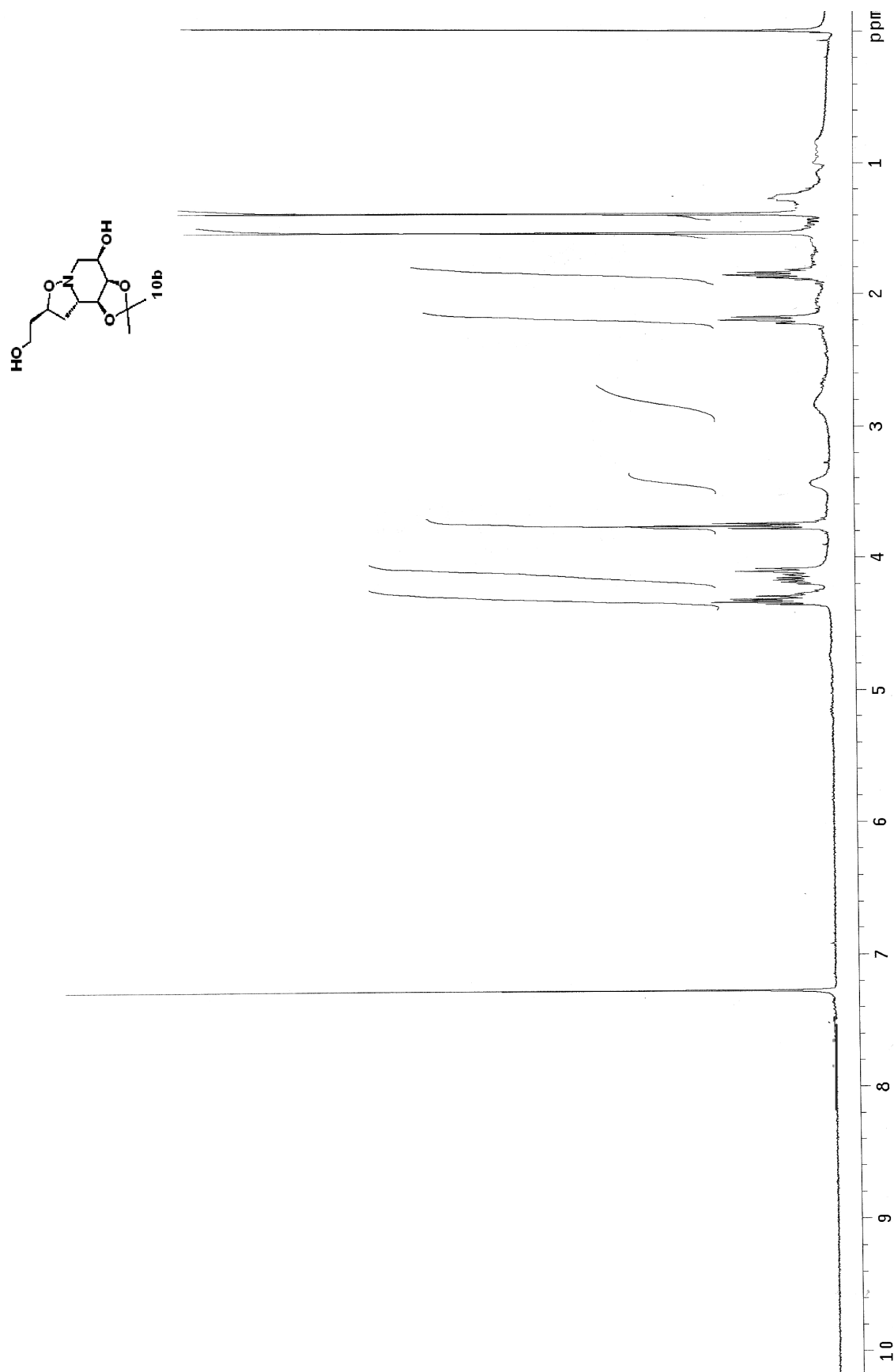


Figure 3S: ^1H (300 MHz, CDCl_3) Spectrum of compound 10b

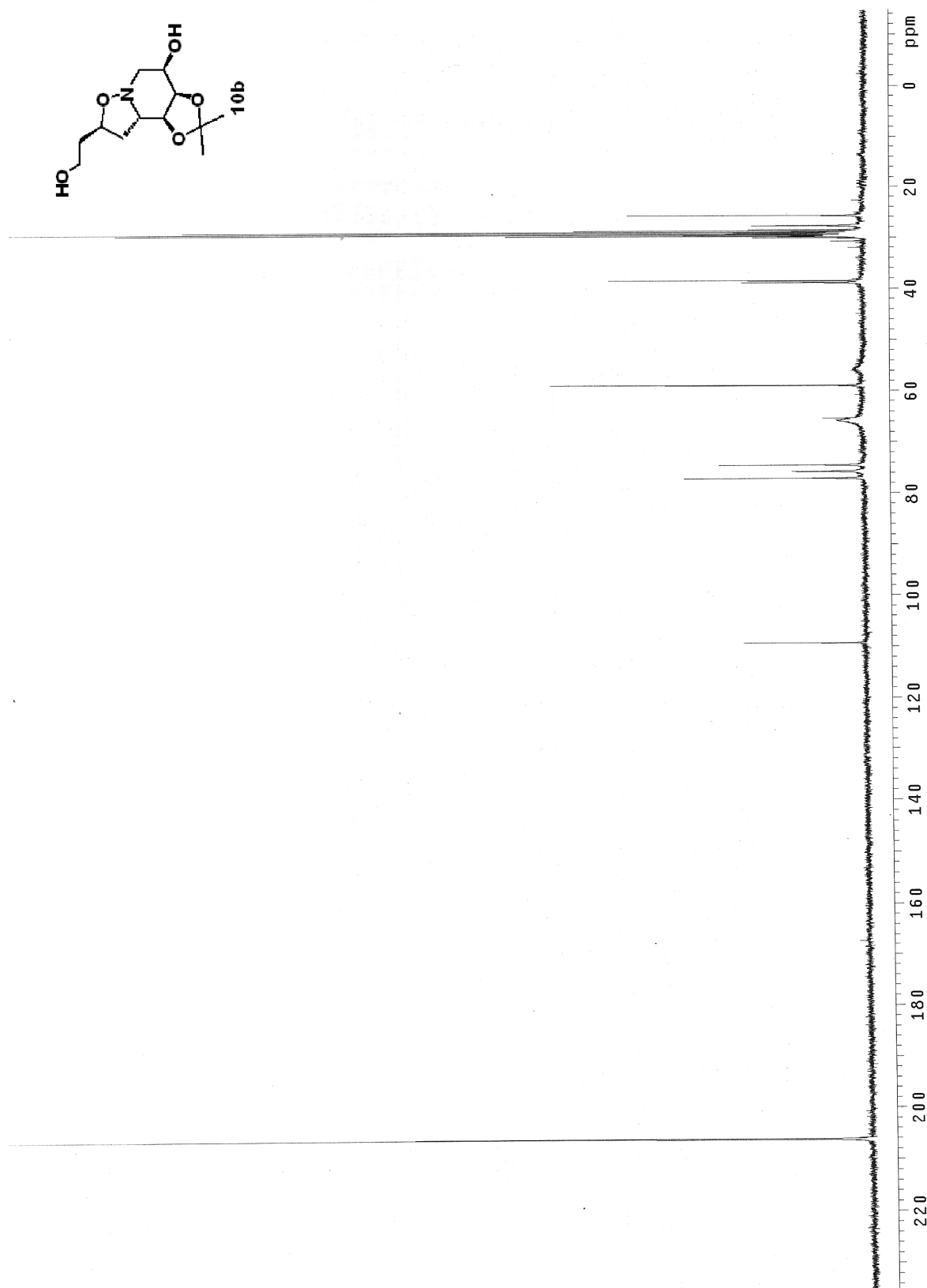
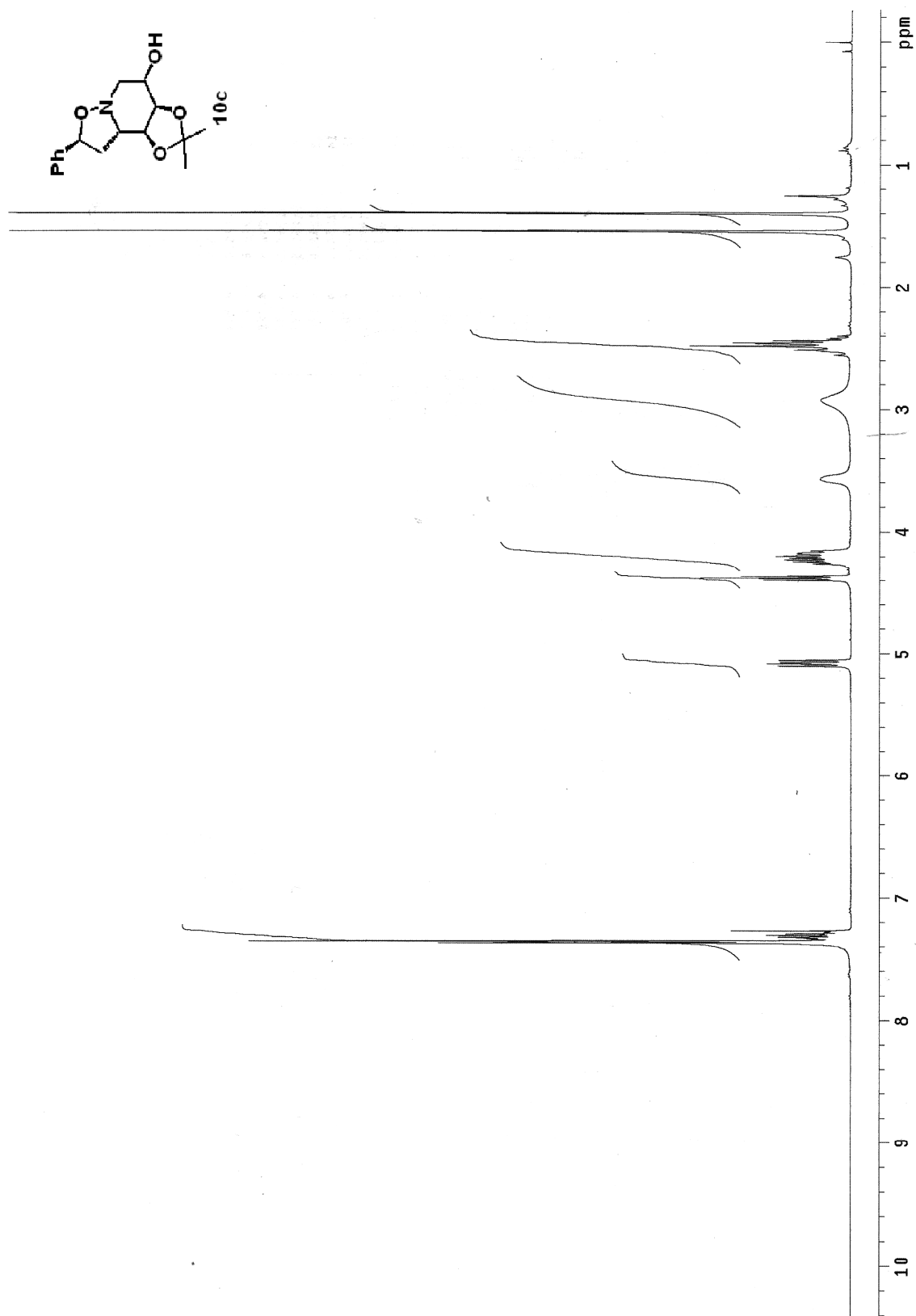


Figure 4S: ^{13}C (75 MHz, Acetone- d_6) Spectrum of compound 10b



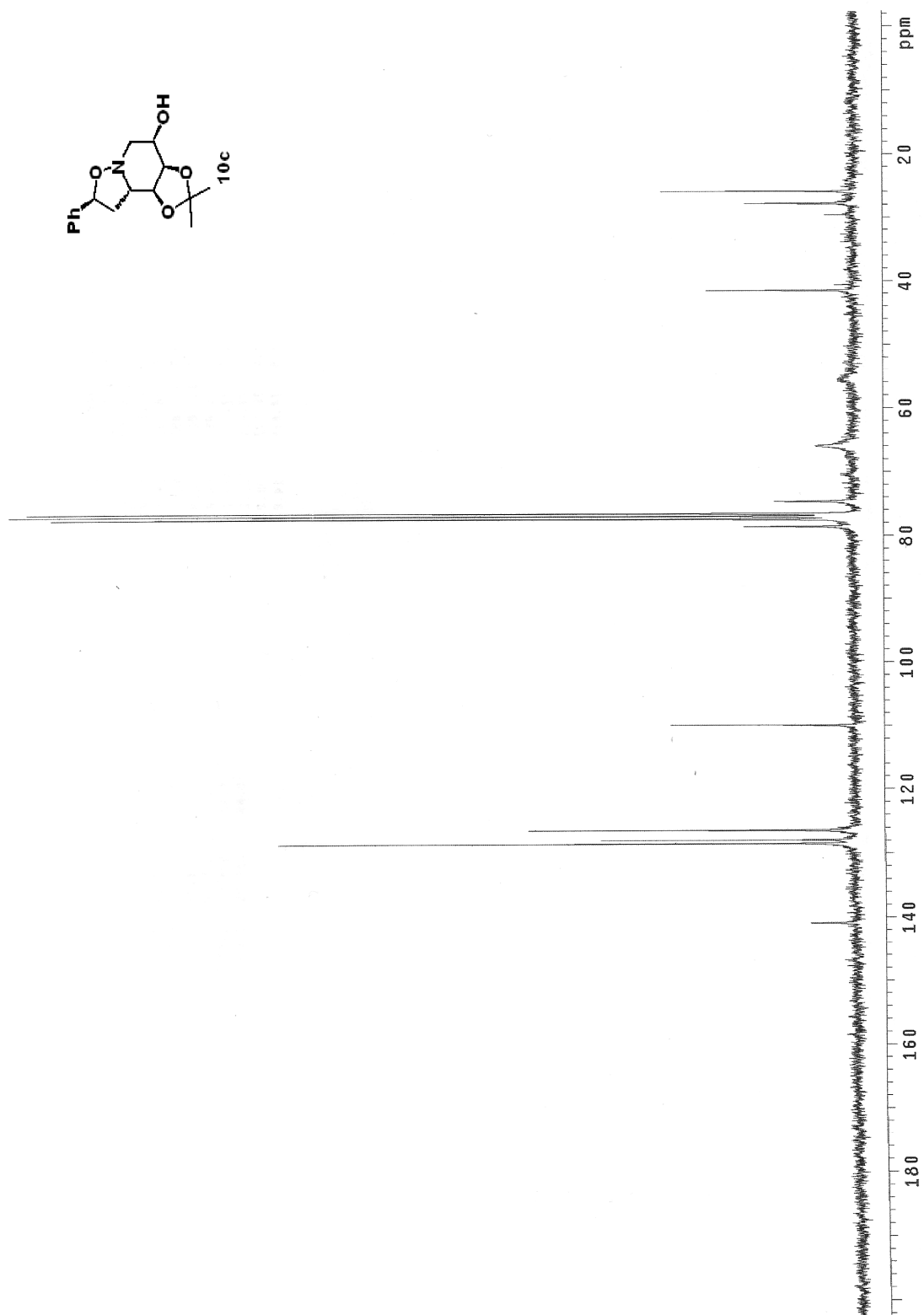


Figure 6S: ^{13}C (75 MHz, CDCl_3) Spectrum of compound 10c

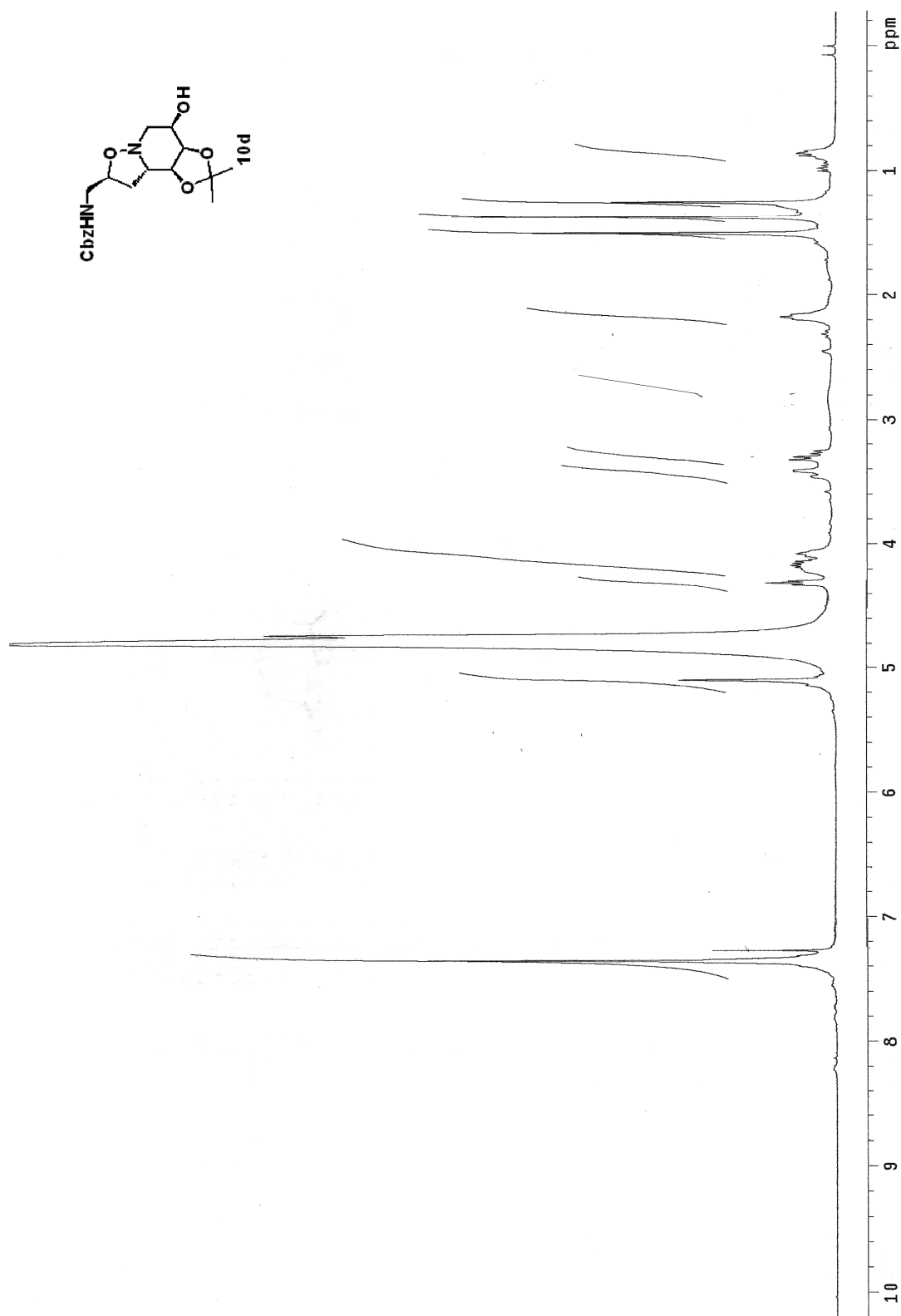


Figure 7S: ^1H (300 MHz, CDCl_3) Spectrum of compound 10d

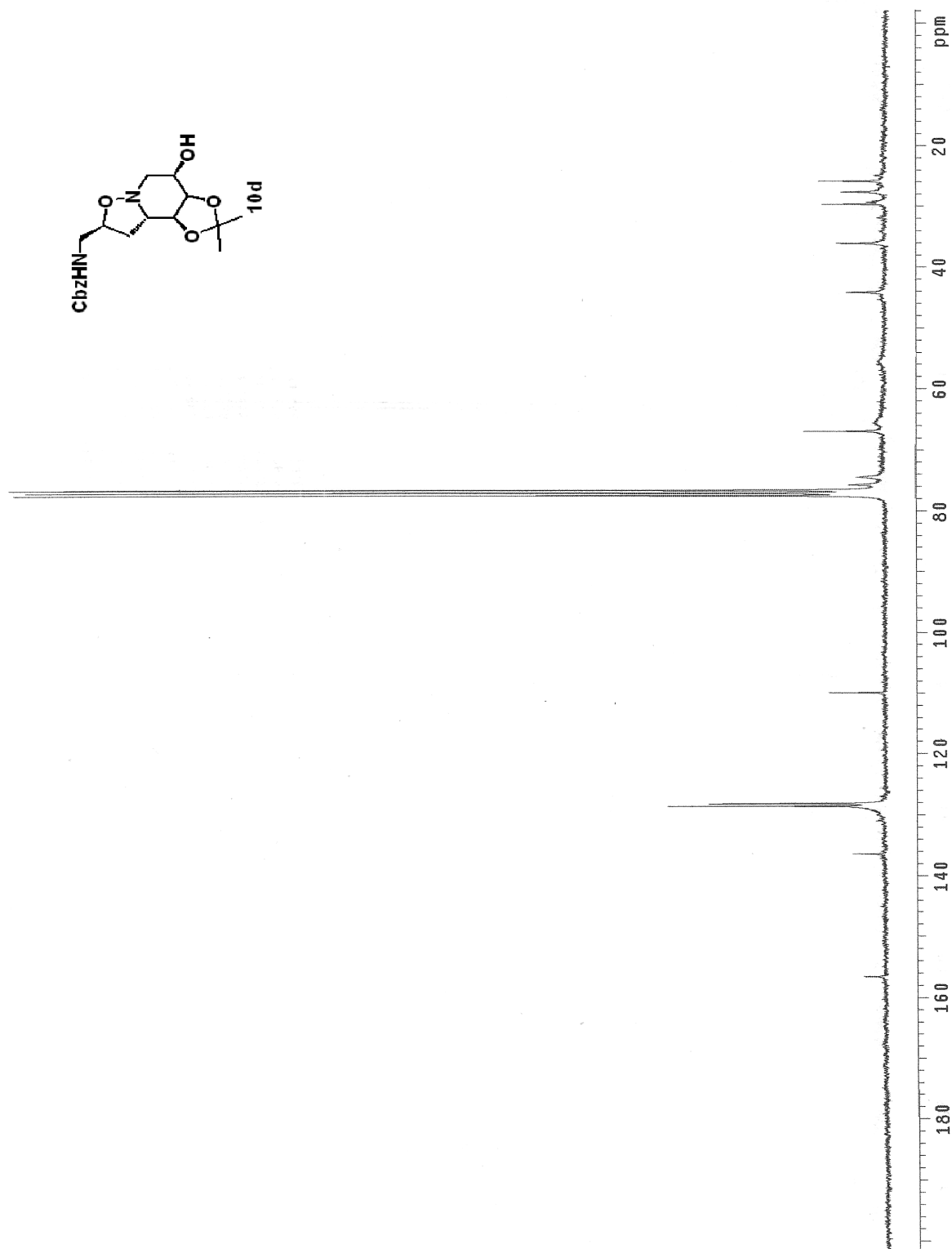


Figure 8S: ^{13}C (75 MHz, CDCl_3) Spectrum of compound 10d

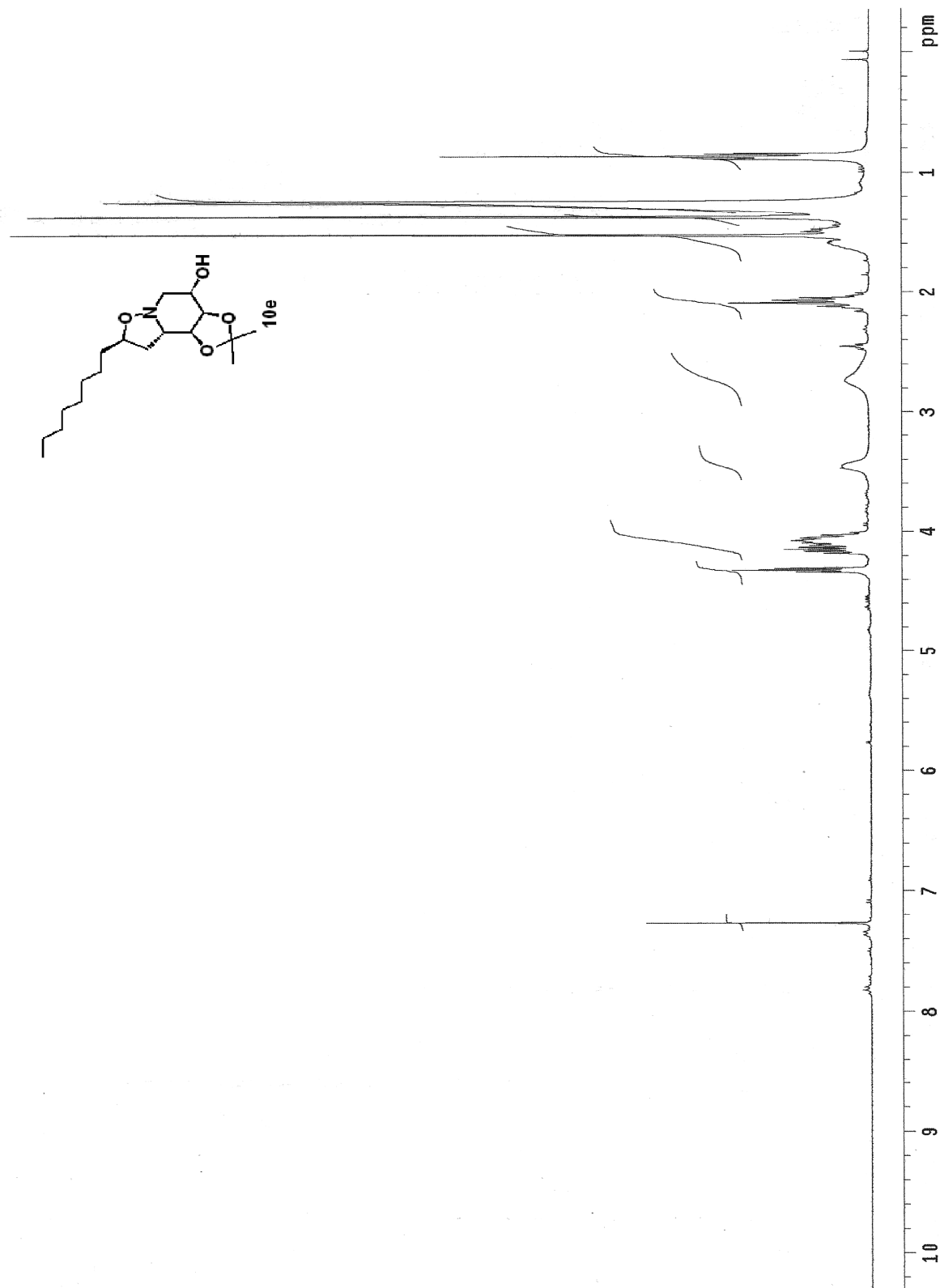


Figure 9S: ^1H (300 MHz, CDCl_3) Spectrum of compound 10e

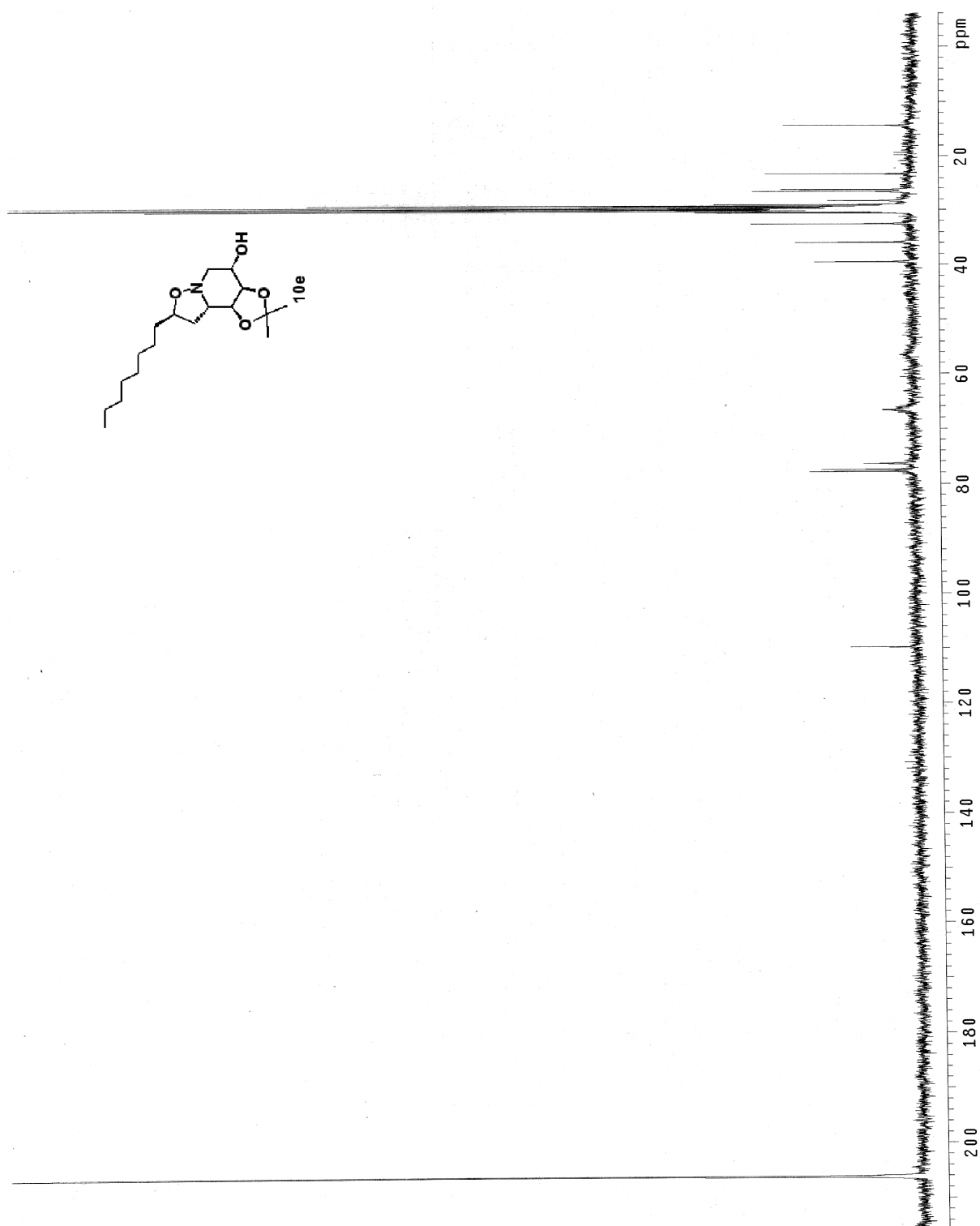


Figure 10S: ^{13}C (75 MHz, Acetone- d_6) Spectrum of compound 10e

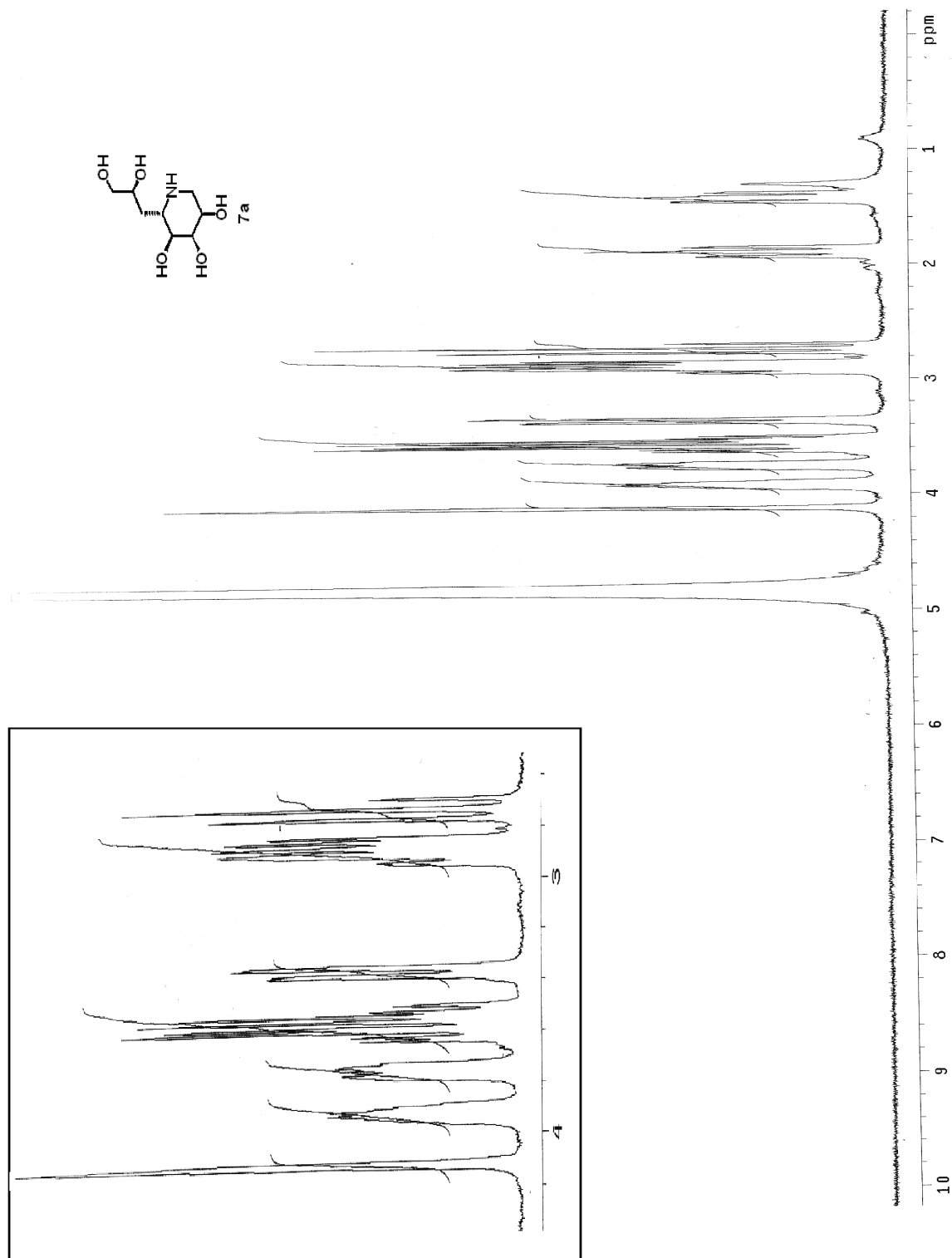


Figure 11S: ^1H (300 MHz, D_2O) Spectrum of compound **7a**

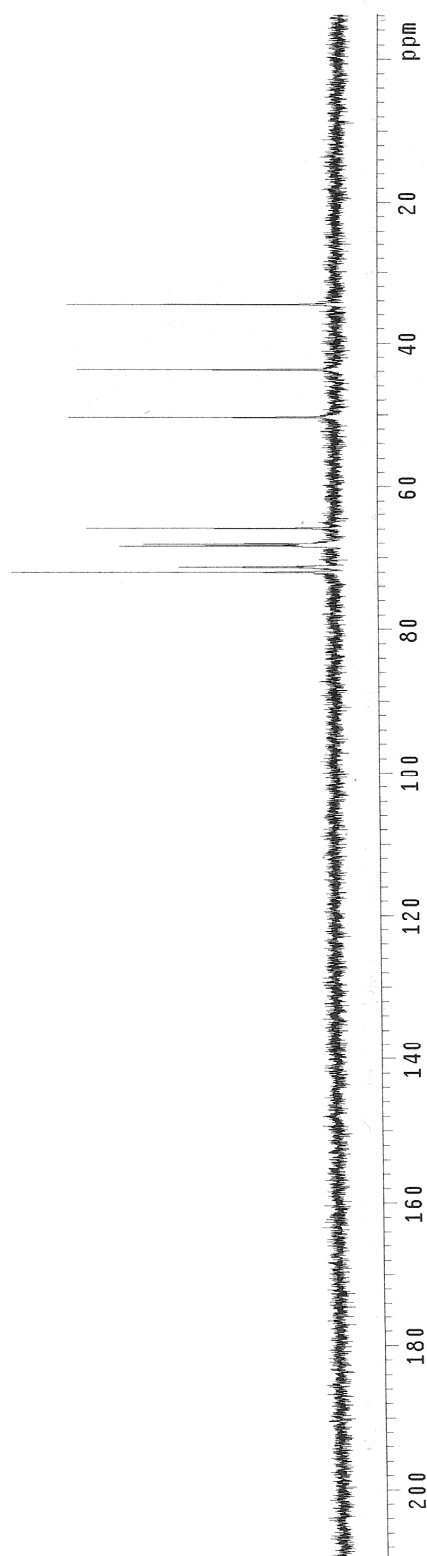
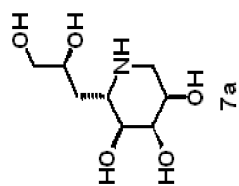


Figure 12S: ^{13}C (75 MHz, D_2O) Spectrum of compound 7a

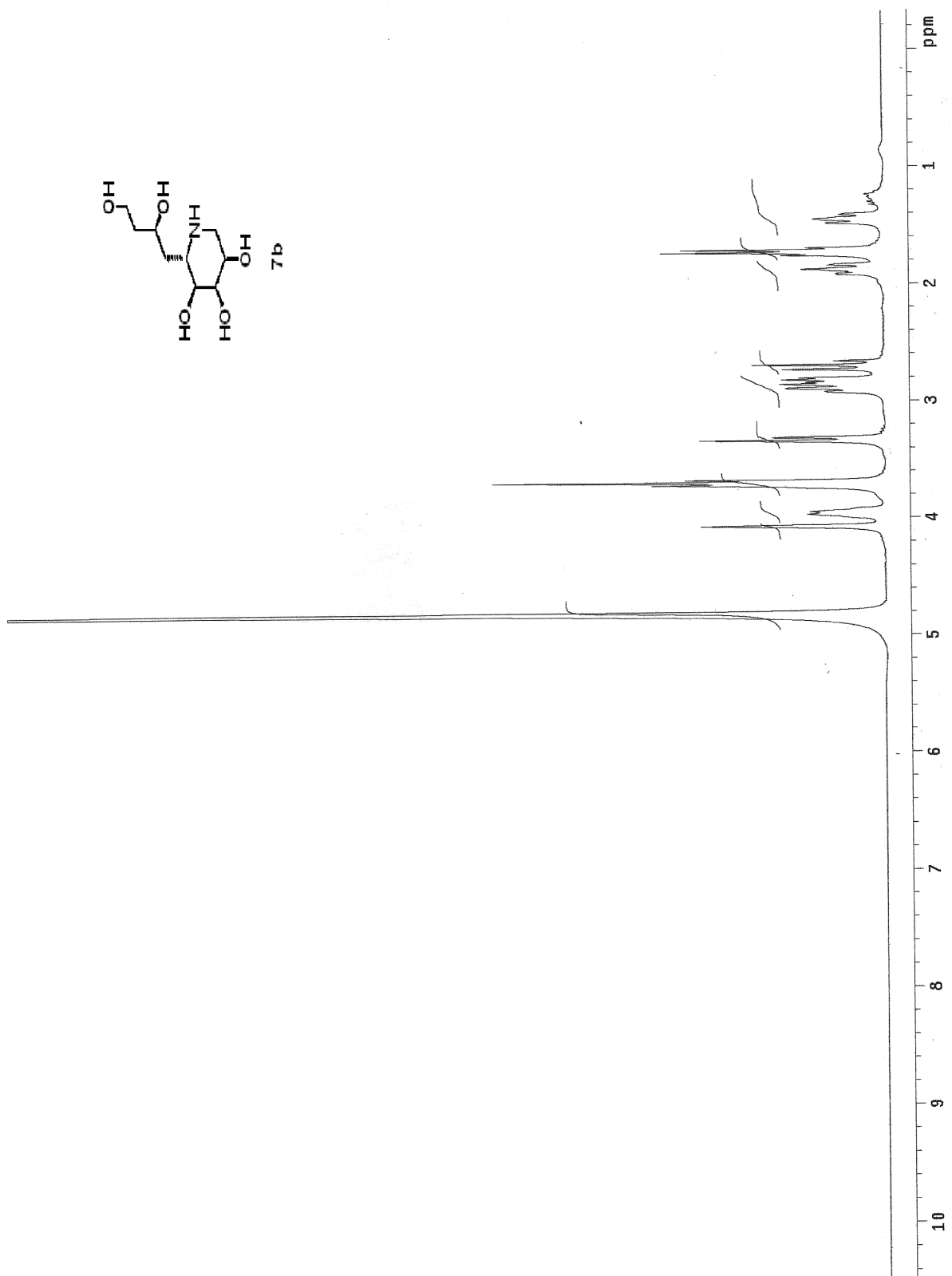


Figure 13S: ¹H (300 MHz, D₂O) Spectrum of compound 7b

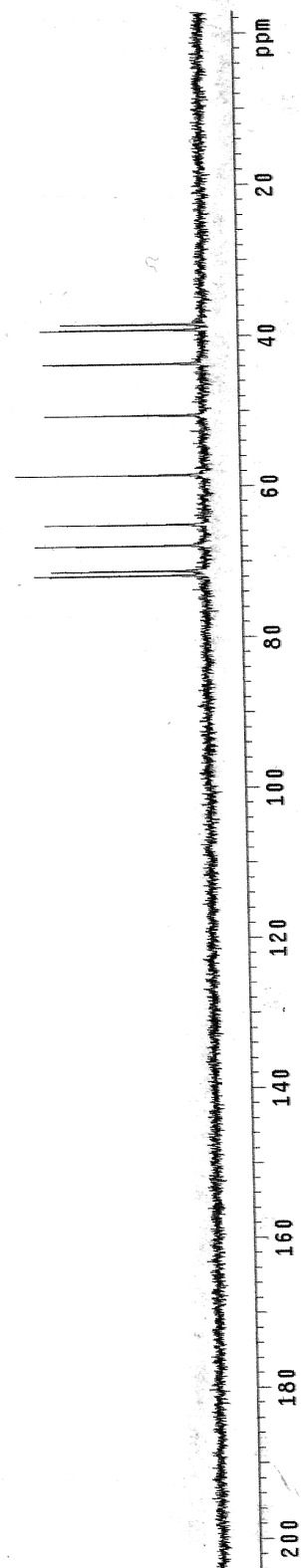
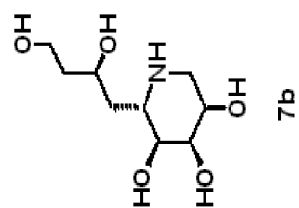


Figure 14S: ^{13}C (75 MHz, D_2O) Spectrum of compound 7b

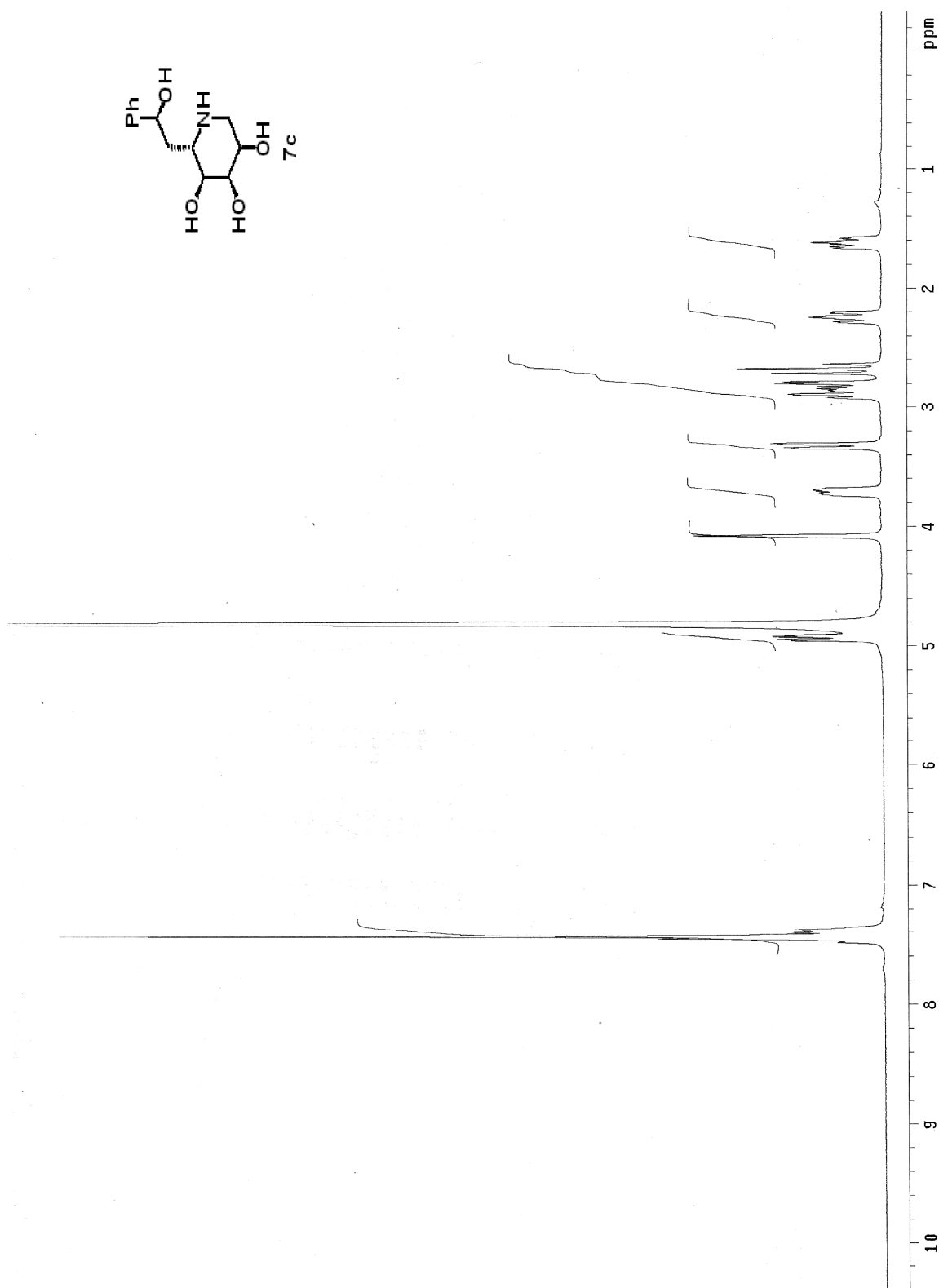


Figure 15S: ^1H (300 MHz, D_2O) Spectrum of compound **7c**

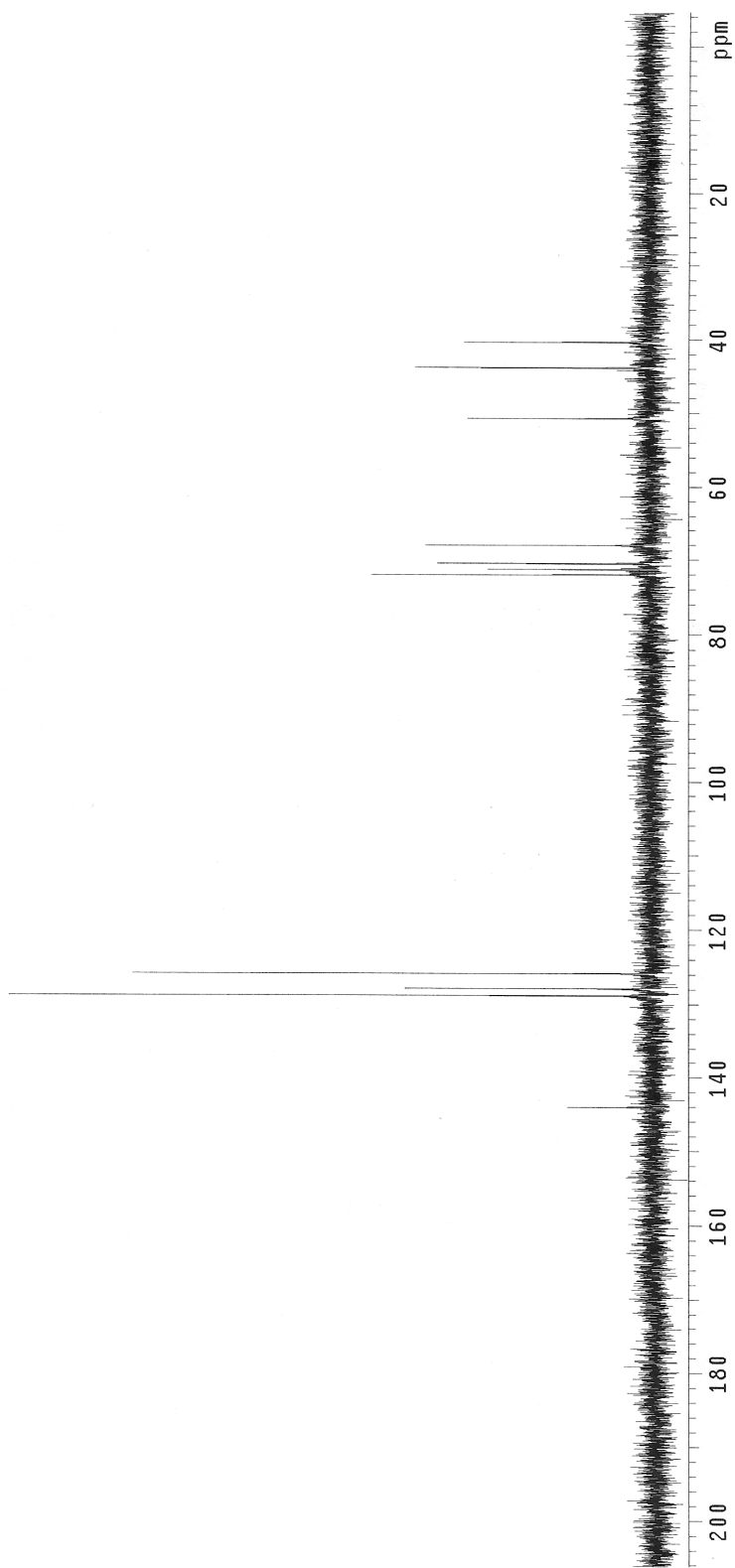
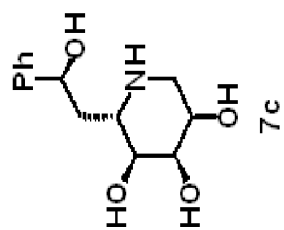


Figure 16S: ^{13}C (75 MHz, D_2O) Spectrum of compound 7c

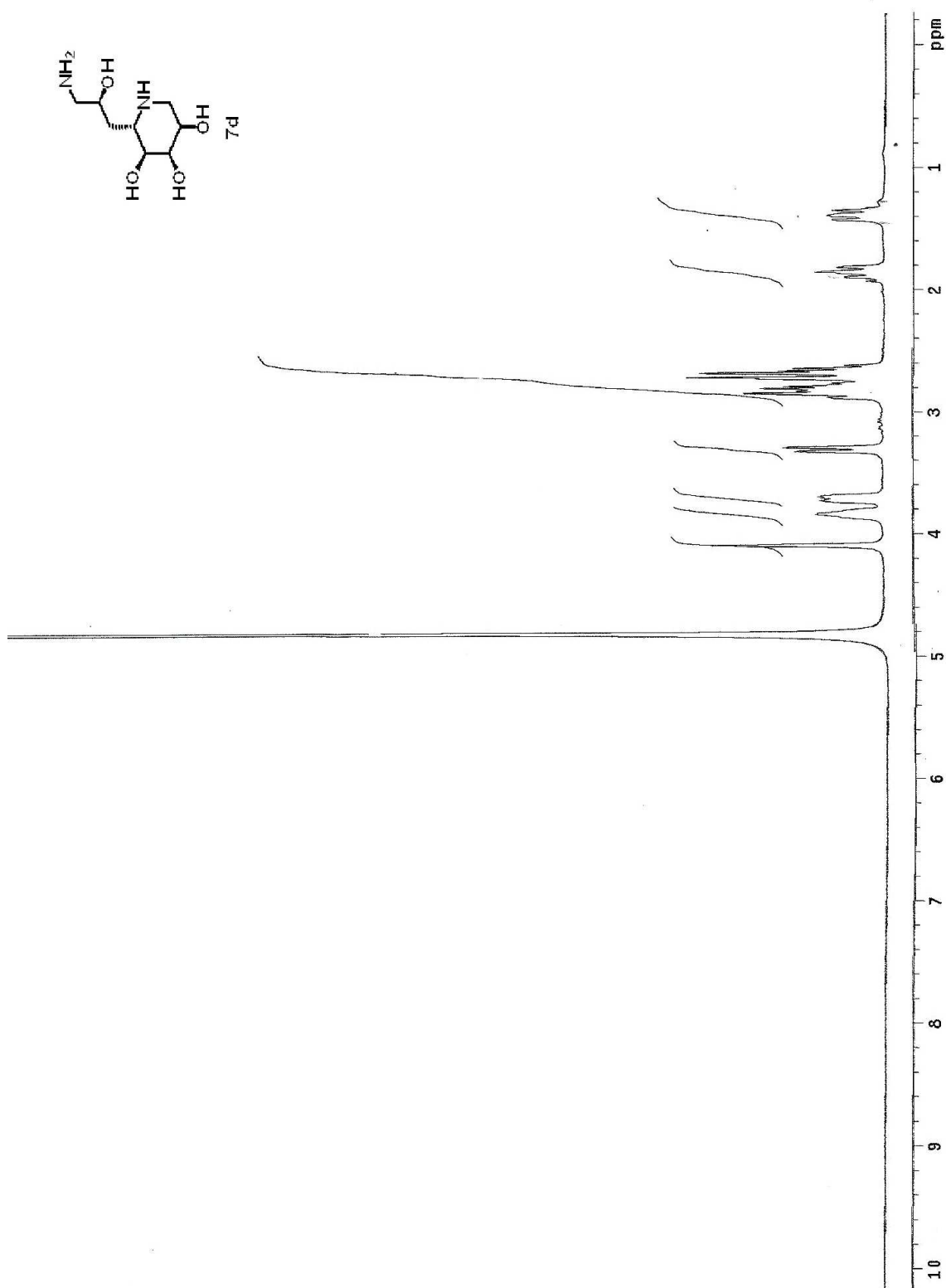


Figure 17S: ^1H (300 MHz, D_2O) Spectrum of compound **7d**

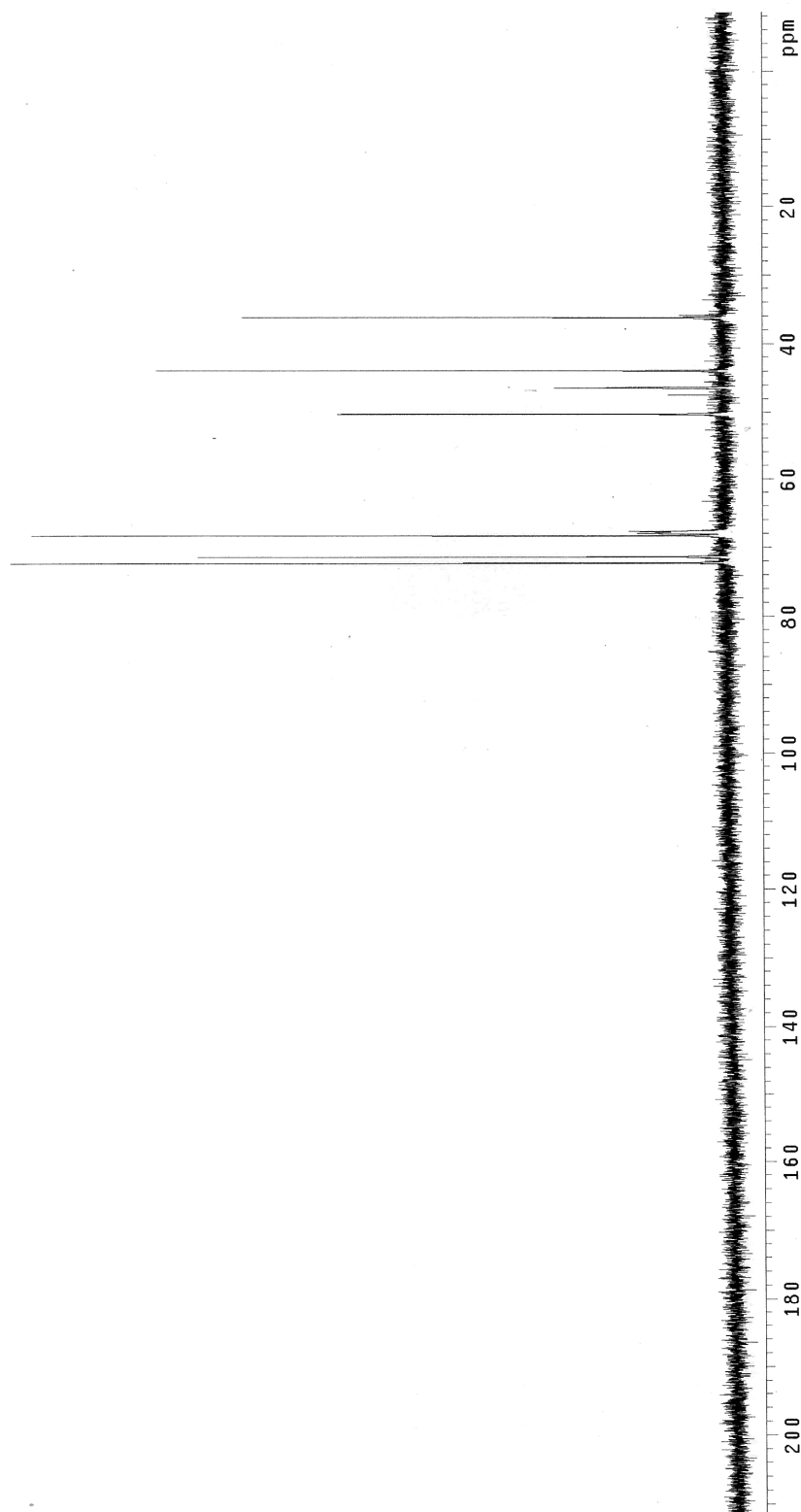
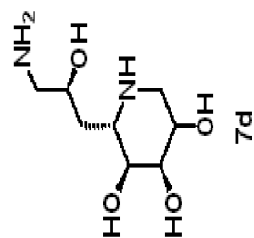


Figure 18S: ^{13}C (75 MHz, D_2O) Spectrum of compound 7d

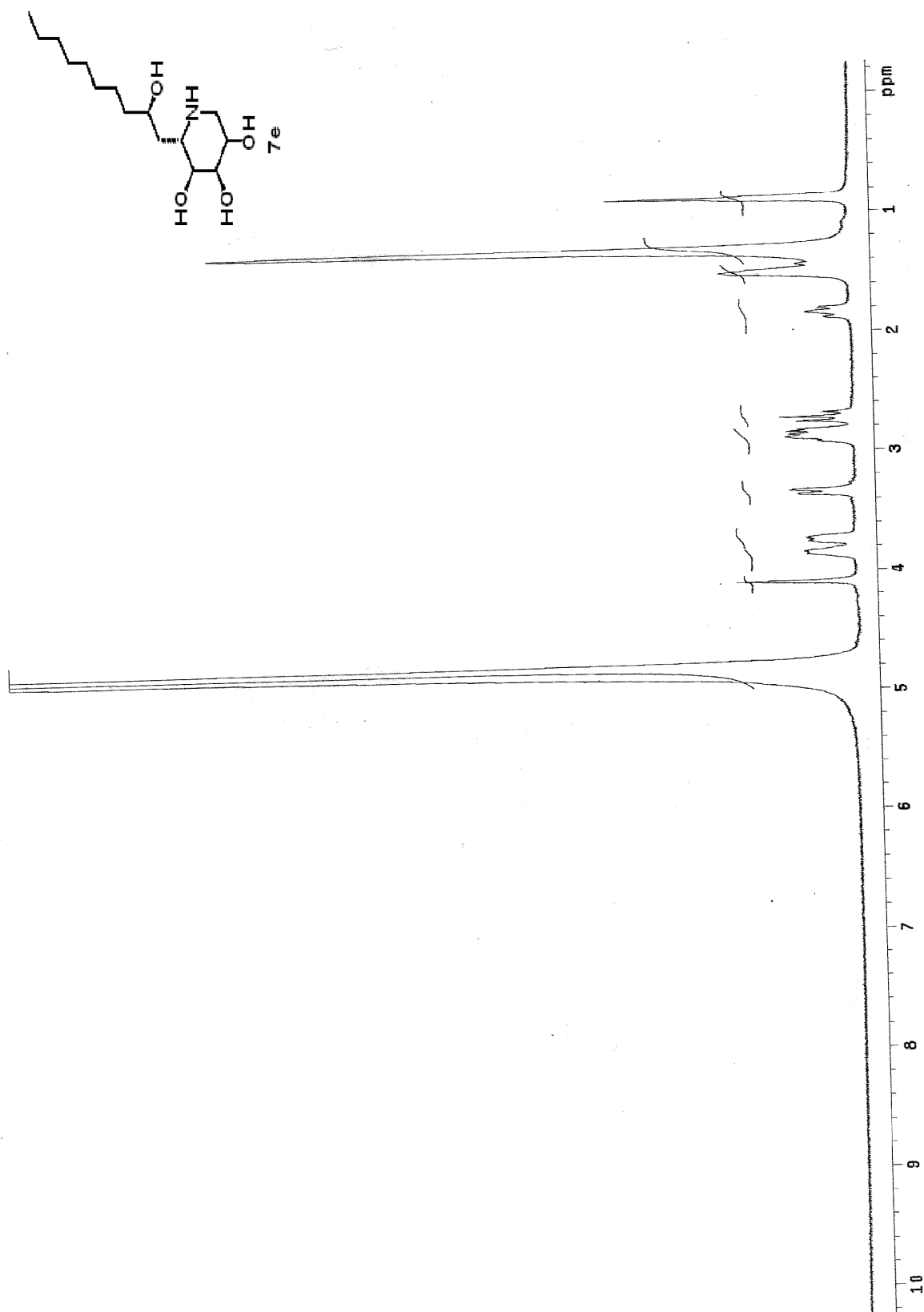


Figure 19S: ^1H (300 MHz, D_2O) Spectrum of compound **7e**

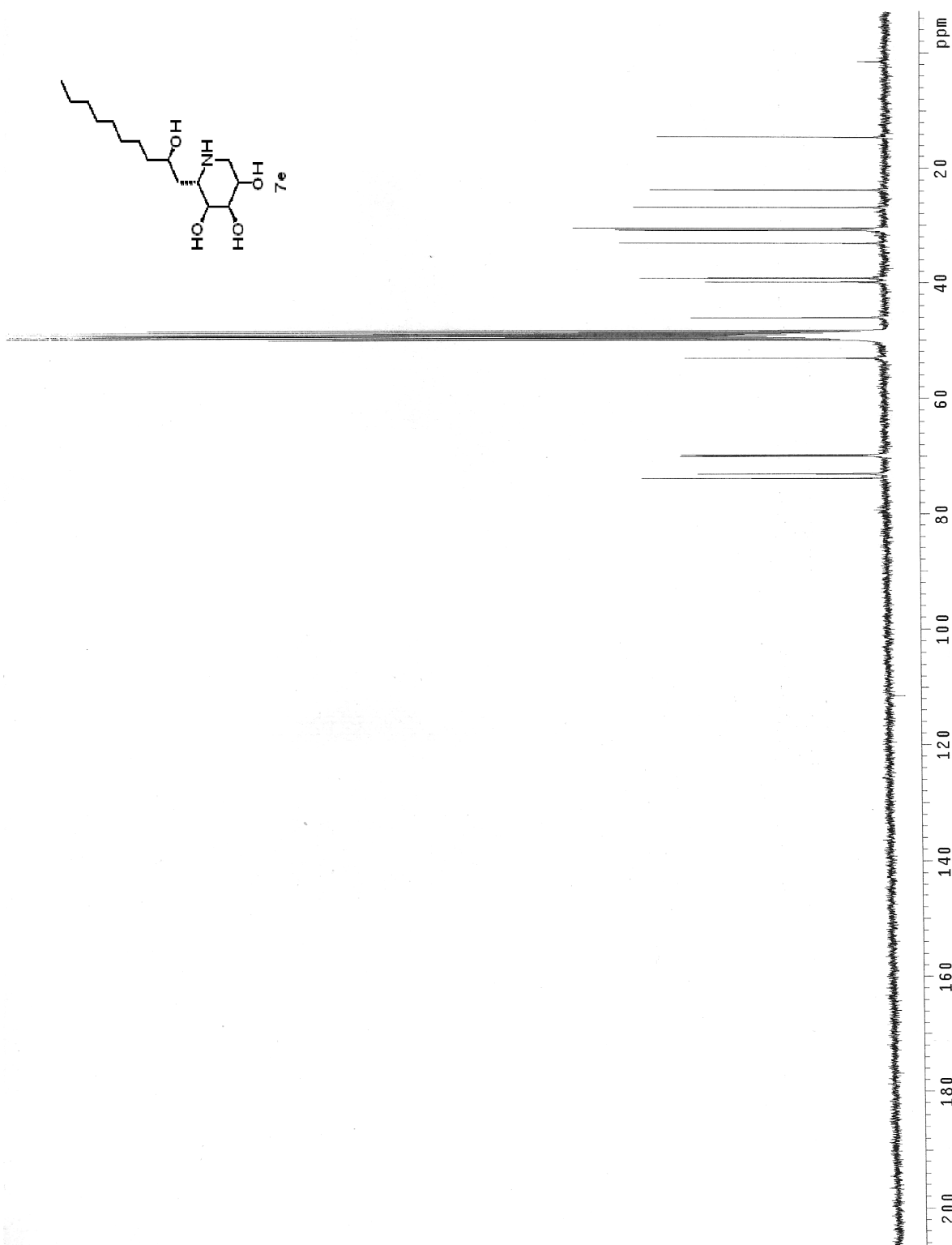


Figure 20S: ^{13}C (75 MHz, D_2O) Spectrum of compound **7e**

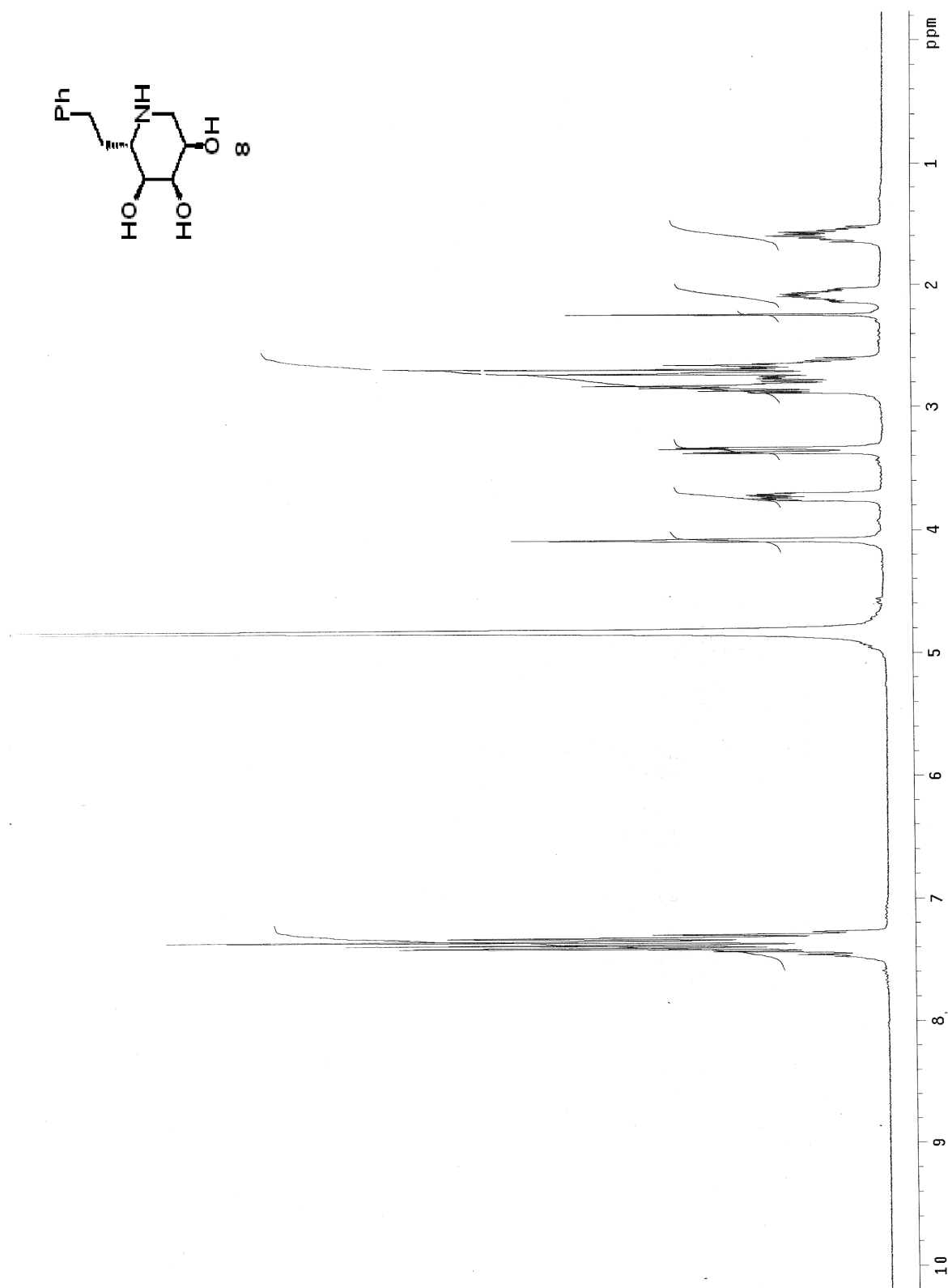


Figure 21S: ¹H (300 MHz, D₂O) Spectrum of compound 8

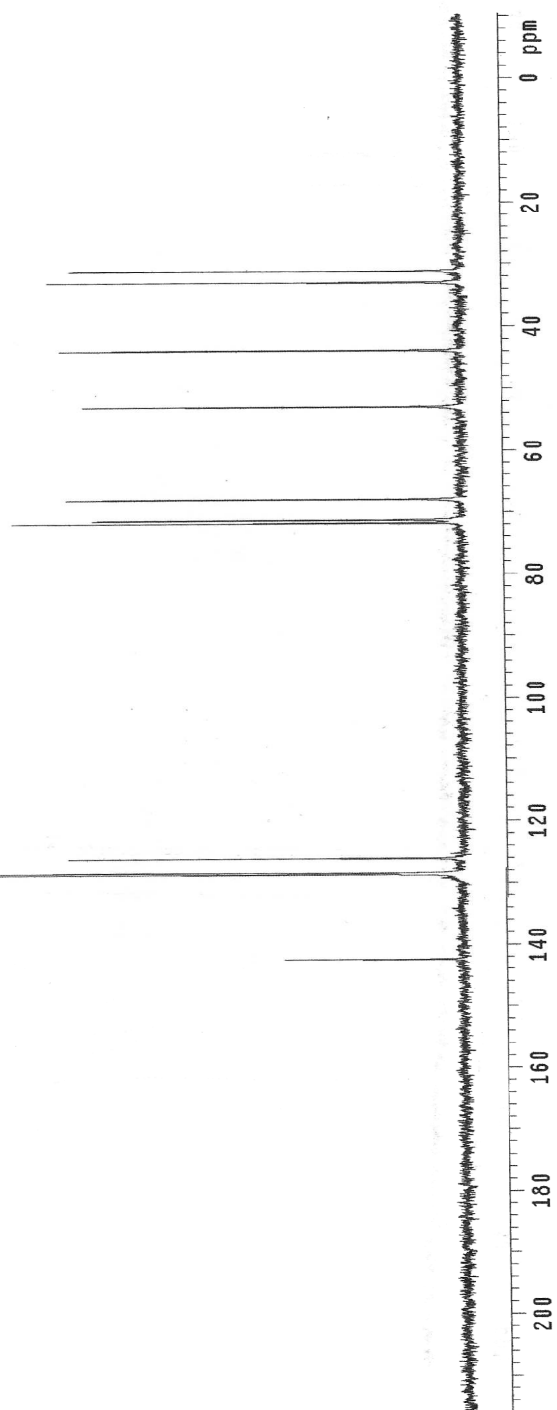
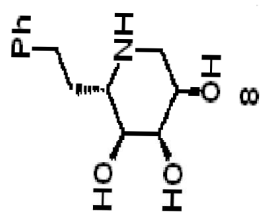


Figure 22S: ^{13}C (75 MHz, D_2O) Spectrum of compound 8

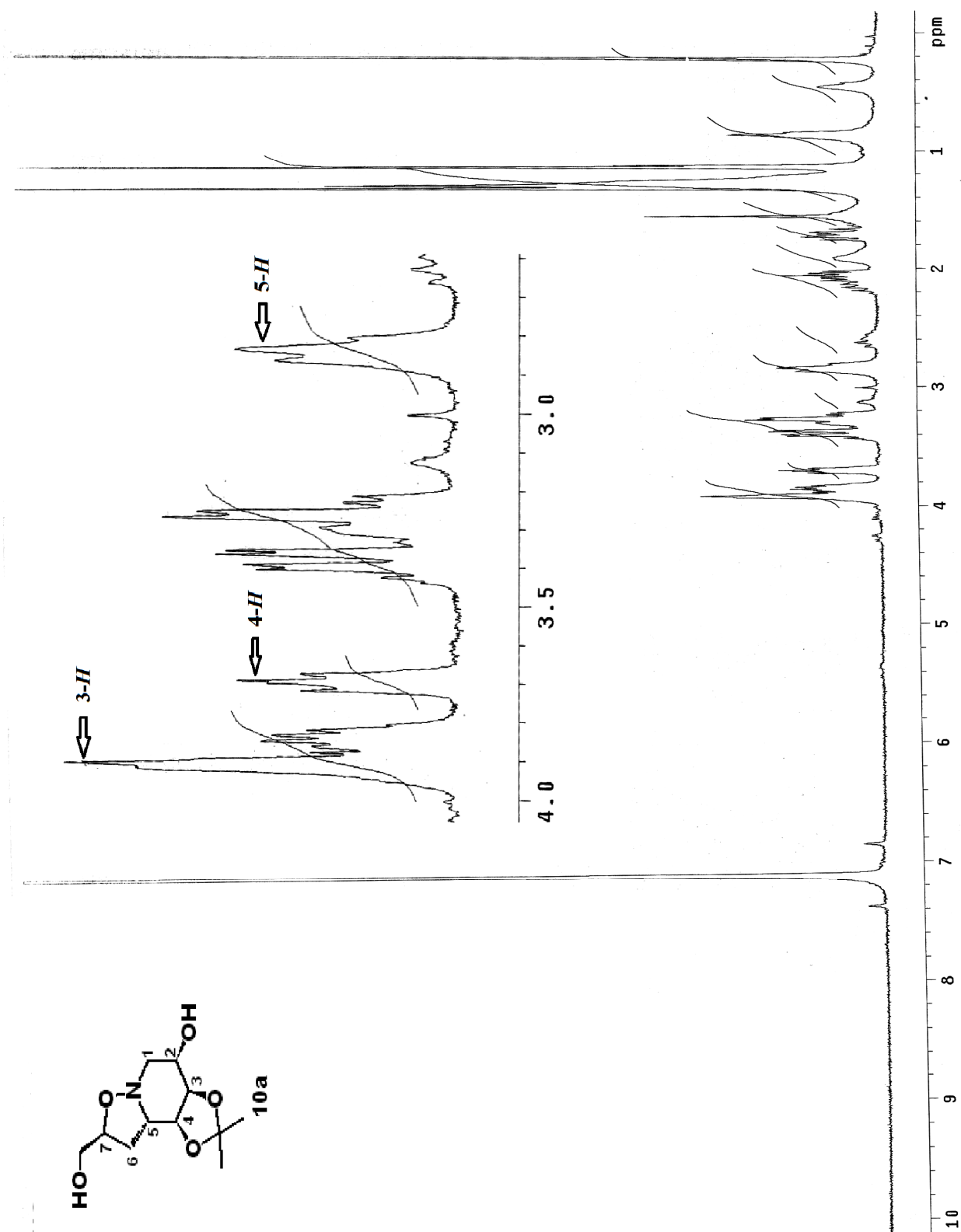


Figure 23S: ^1H (300 MHz, benzene- d_6) Spectrum of compound 10a at 50°C

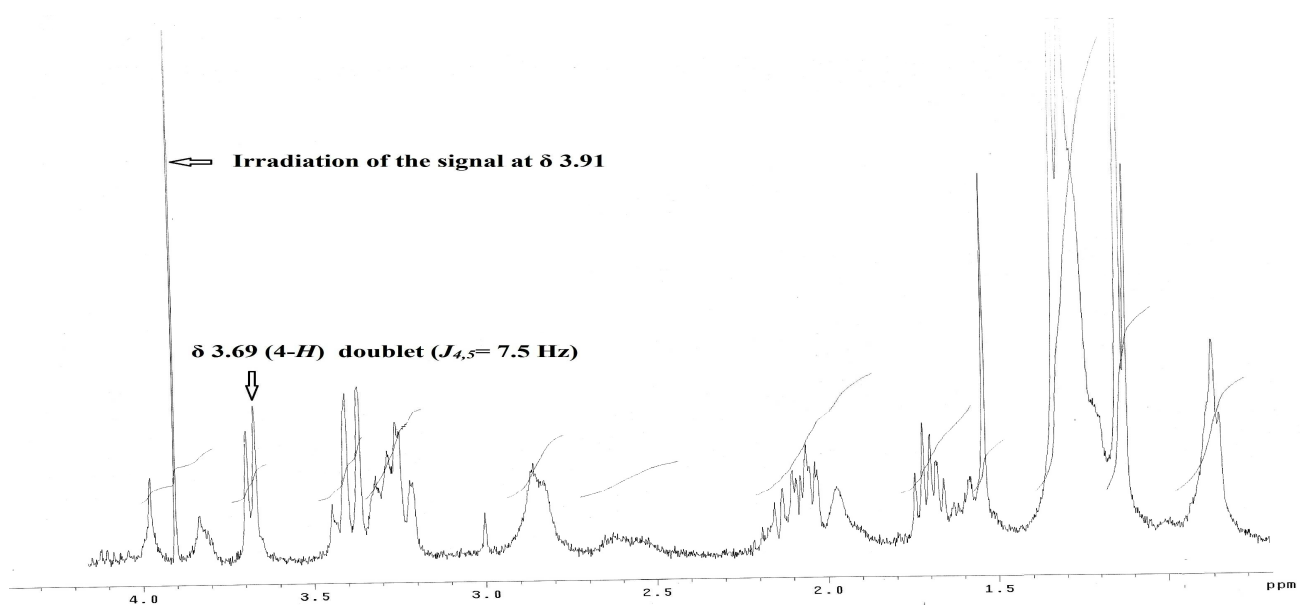


Figure 23.1S: ^1H (300 MHz, benzene- d_6) Decoupled Spectrum of compound 10a at 50°C

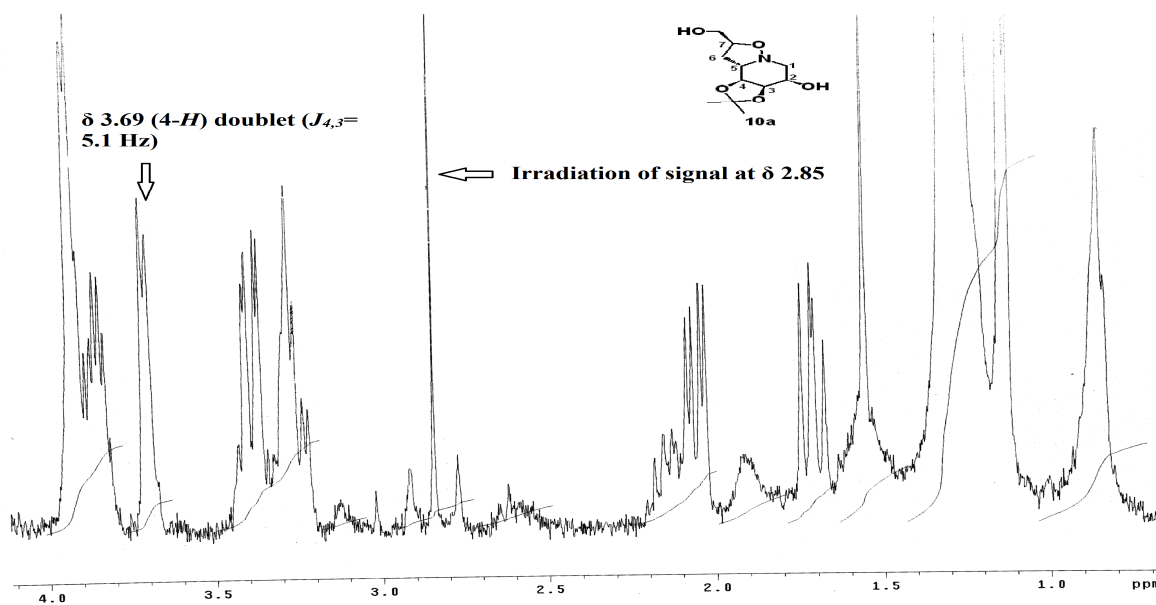
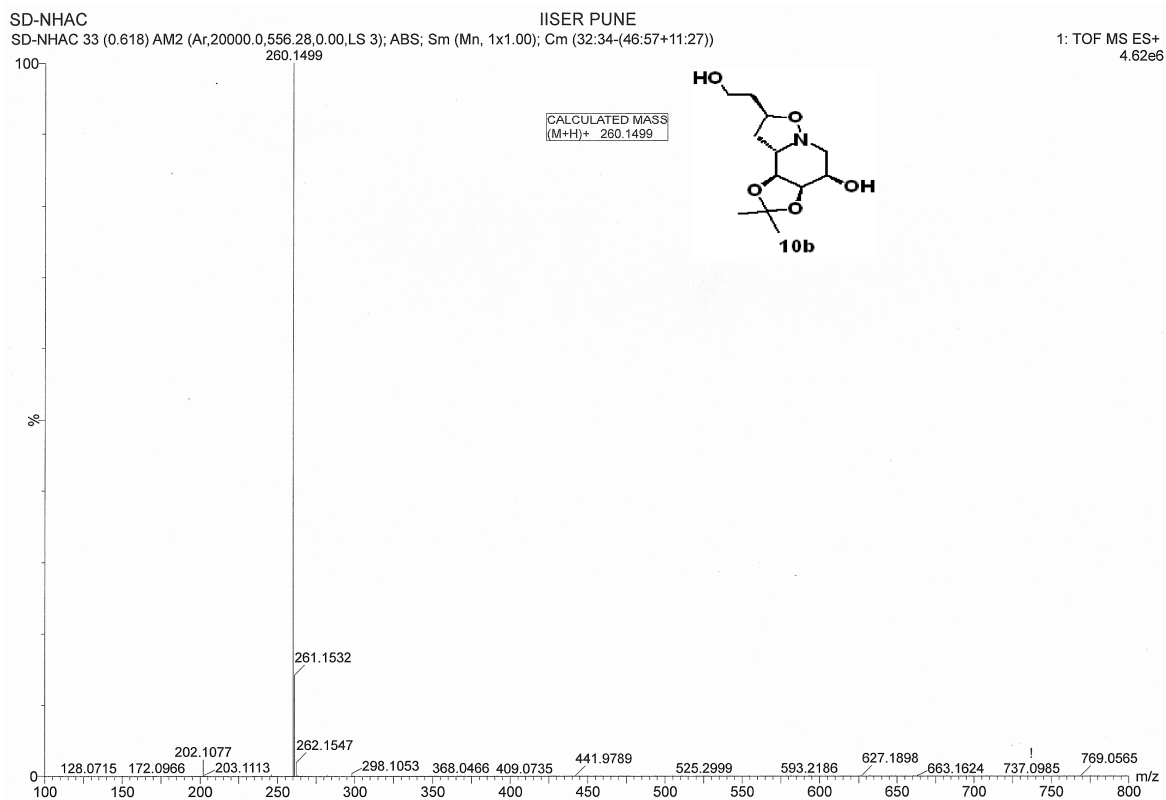
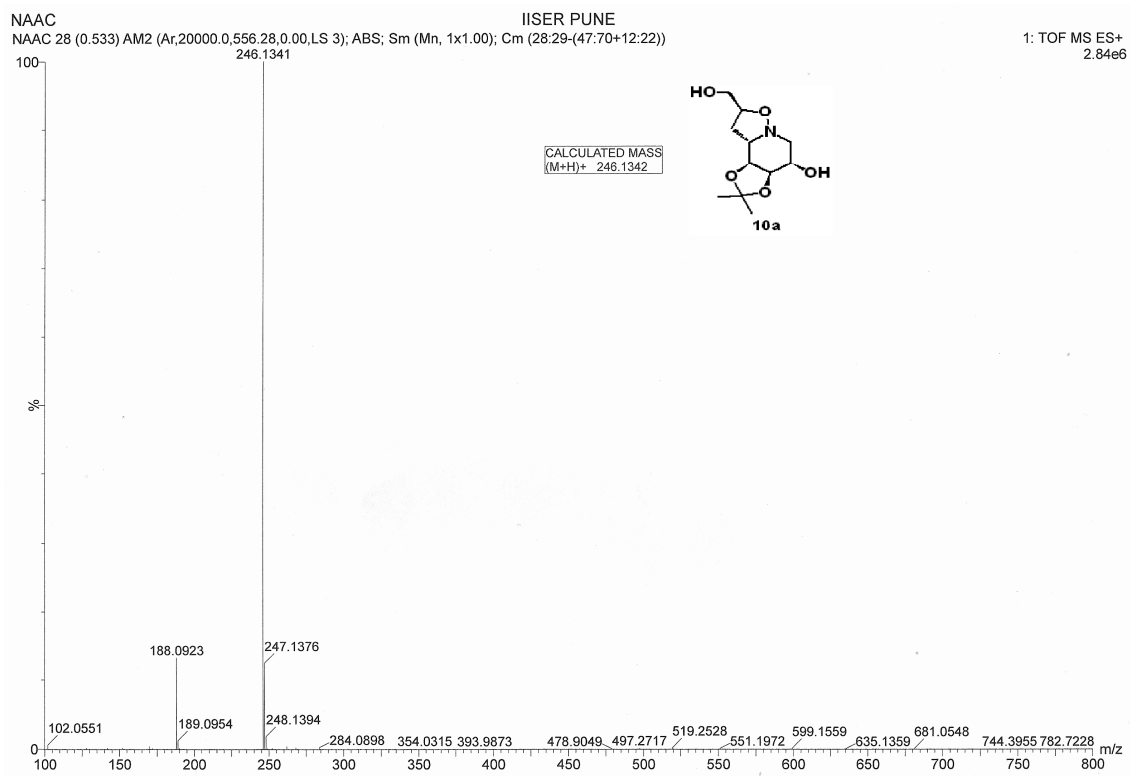


Figure 23.2S: ^1H (300 MHz, benzene- d_6) Decoupling Spectrum of compound 10a at 50°C

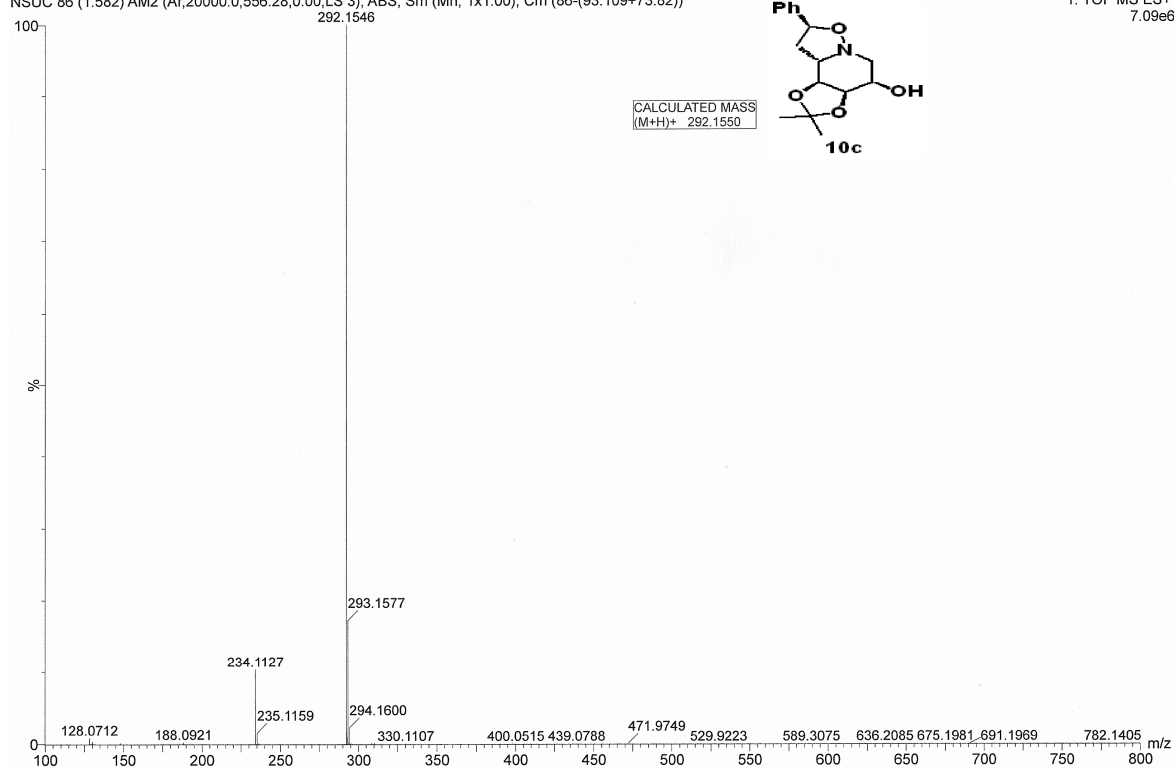


NSUC

IISER PUNE

NSUC 86 (1.582) AM2 (Ar,20000.0,556.28,0.00,LS 3); ABS; Sm (Mn, 1x1.00); Cm (86-(93:109+73:82))

1: TOF MS ES+
7.09e6

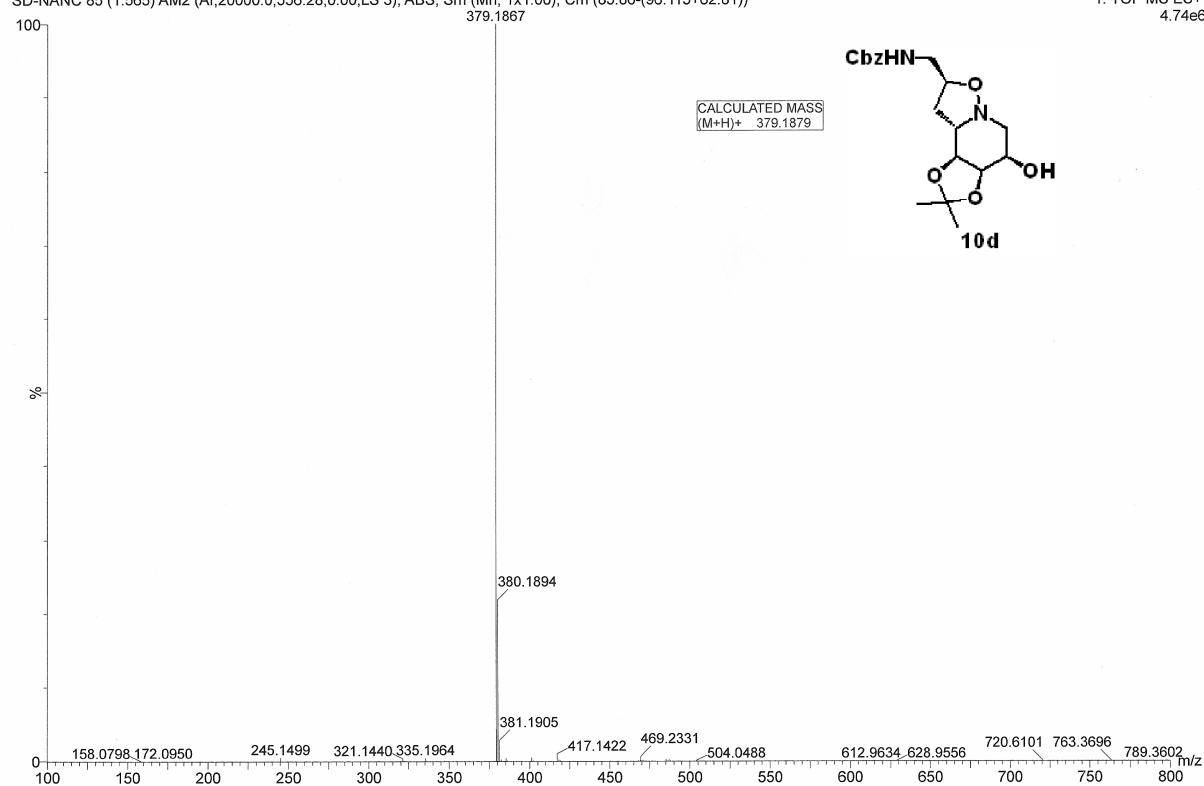


SD-NANC

IISER PUNE

SD-NANC 85 (1.565) AM2 (Ar,20000.0,556.28,0.00,LS 3); ABS; Sm (Mn, 1x1.00); Cm (85:86-(96:115+62:81))

1: TOF MS ES+
4.74e6

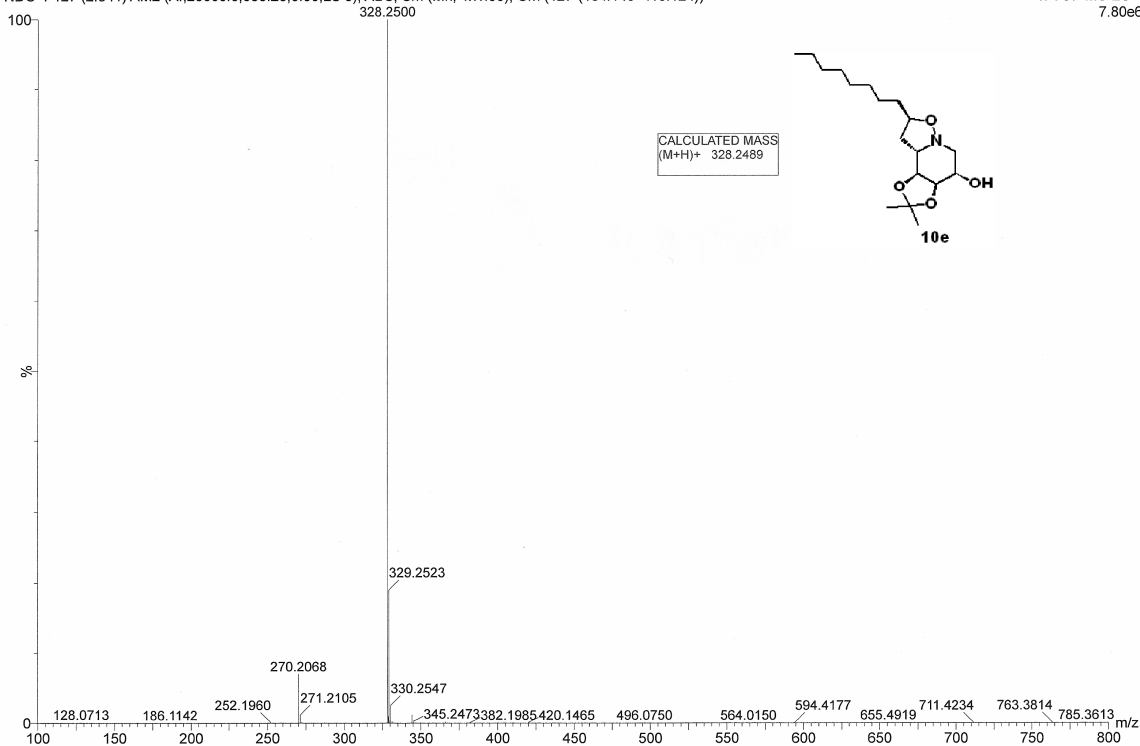


NDC-1

IISER PUNE

NDC-1 127 (2.341) AM2 (Ar,20000.0,556.28,0.00,LS 3); ABS; Sm (Mn, 1x1.00); Cm (127-(134:140+118:124))

1: TOF MS ES+
7.80e6

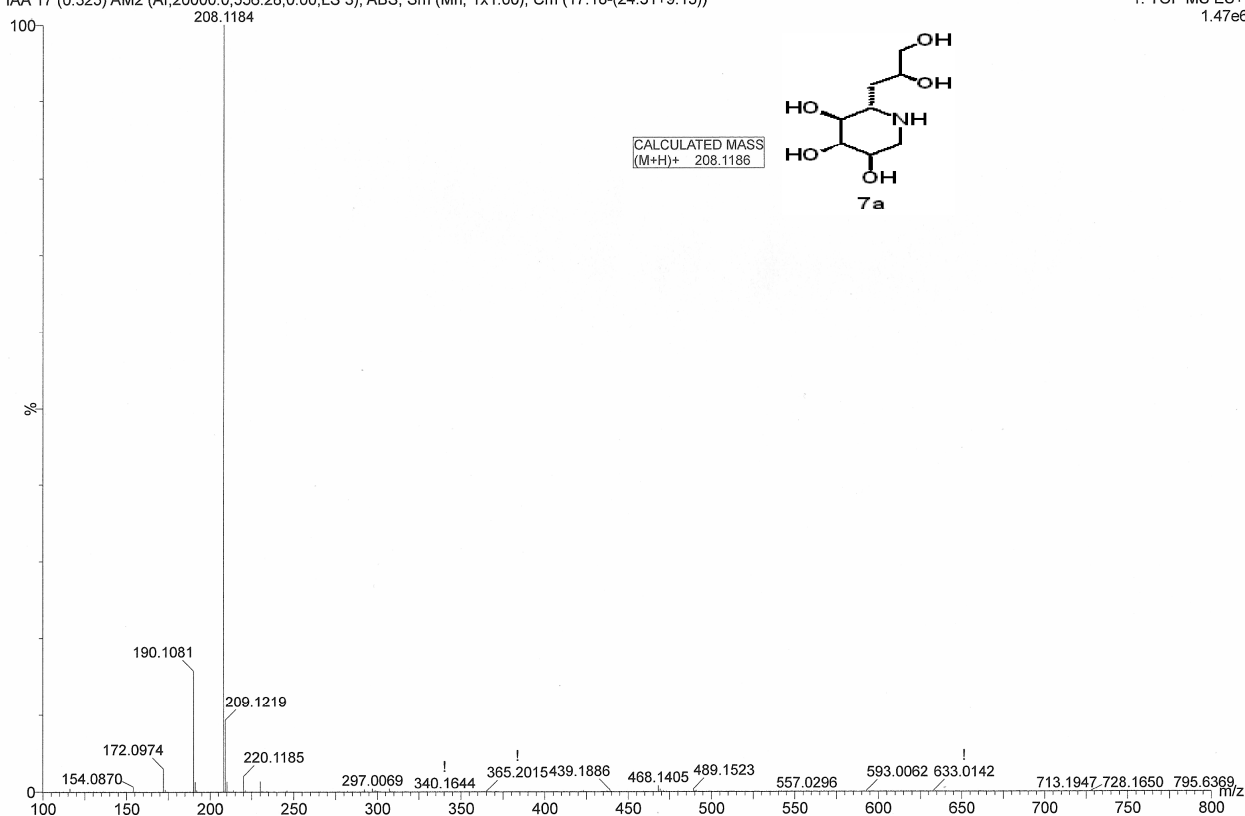


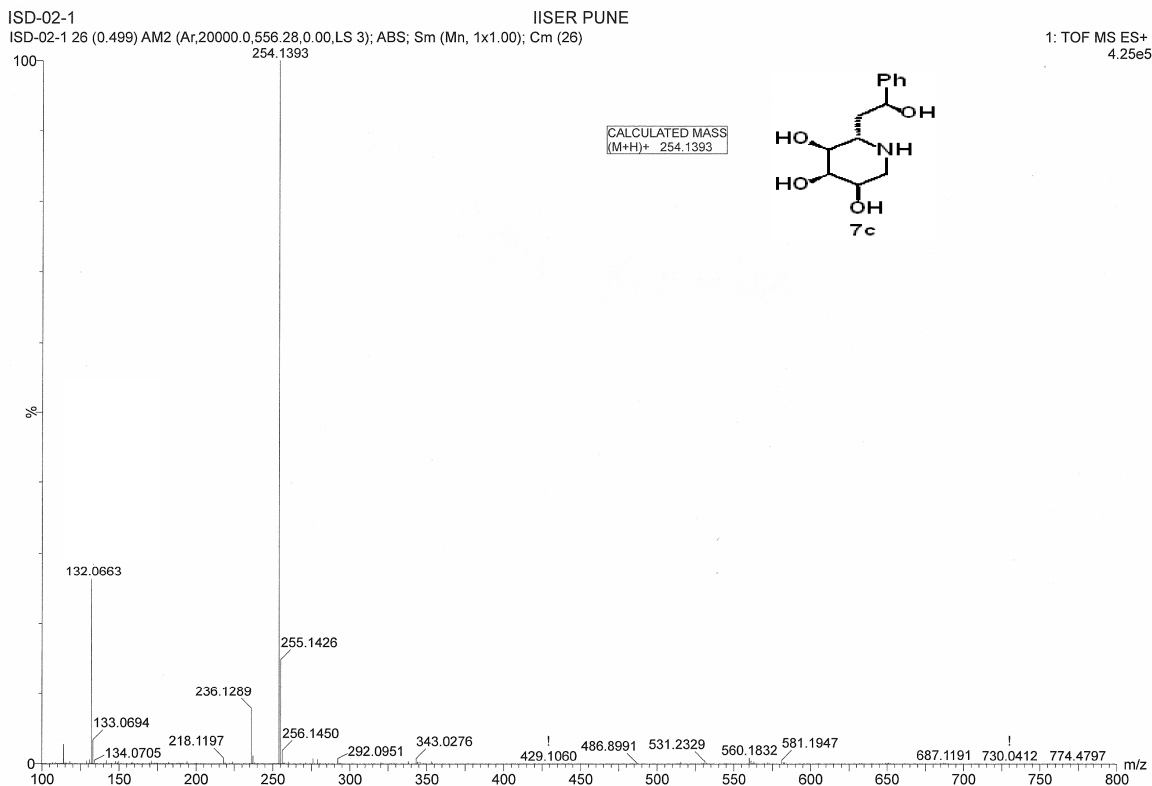
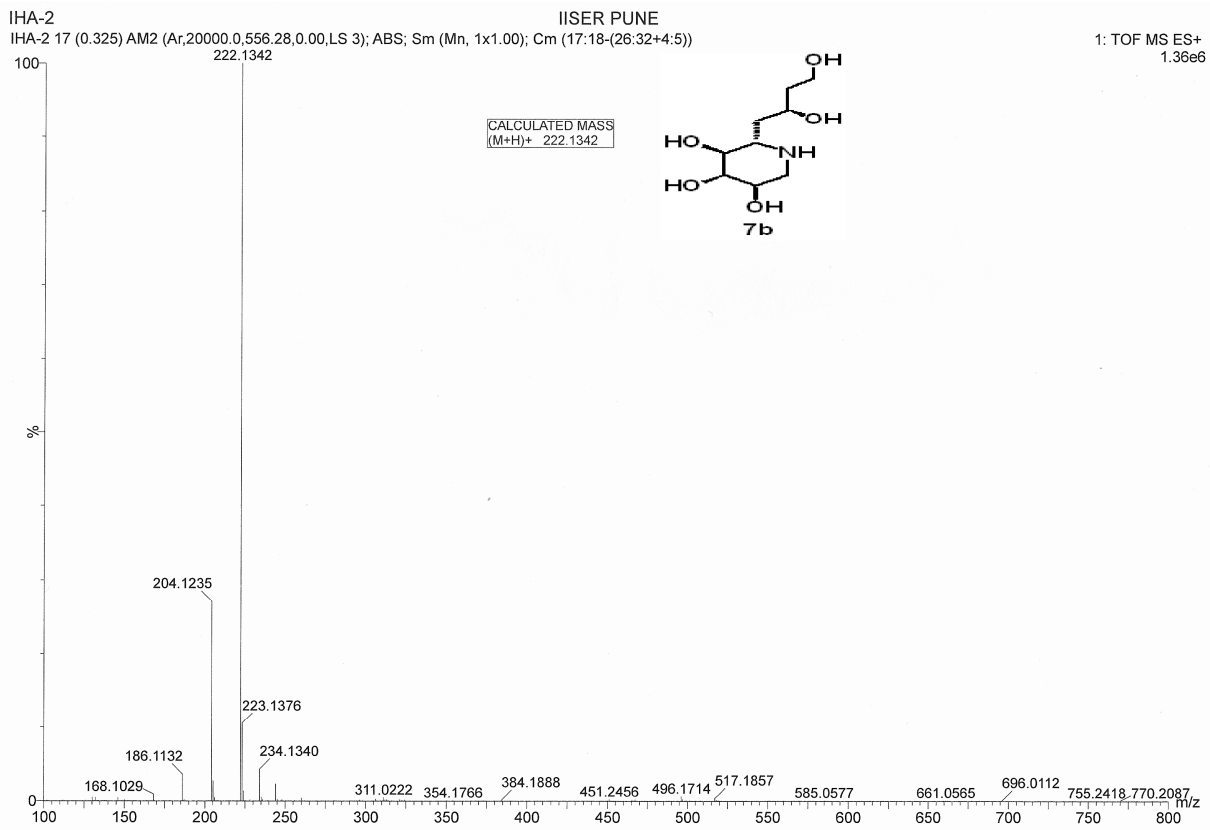
IAA

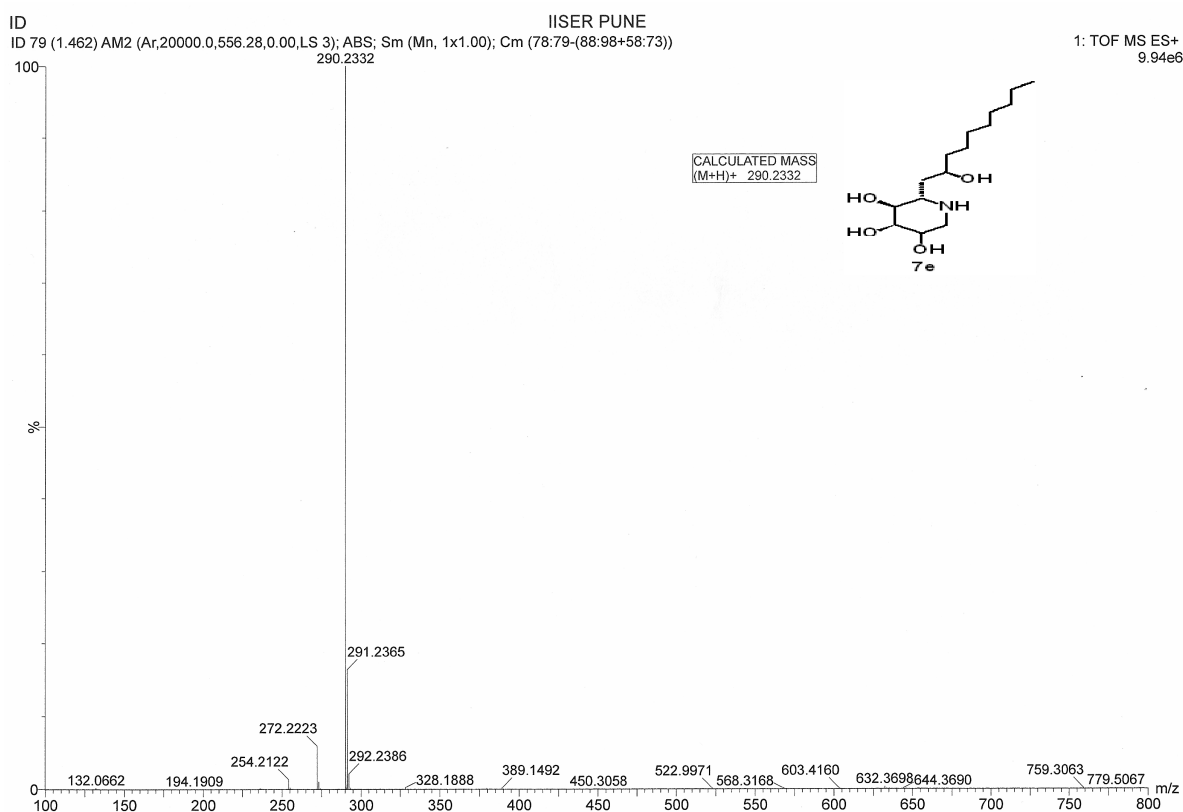
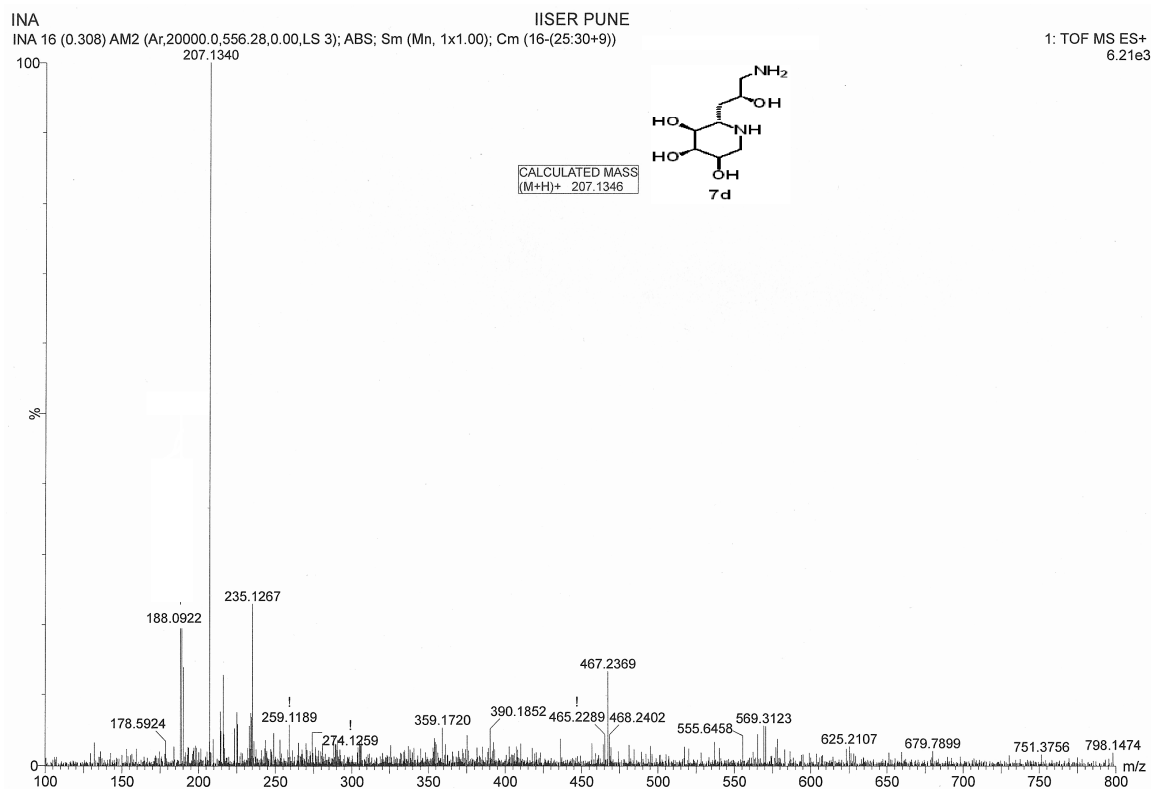
IISER PUNE

IAA 17 (0.325) AM2 (Ar,20000.0,556.28,0.00,LS 3); ABS; Sm (Mn, 1x1.00); Cm (17:18-(24:31+9:13))

1: TOF MS ES+
1.47e6





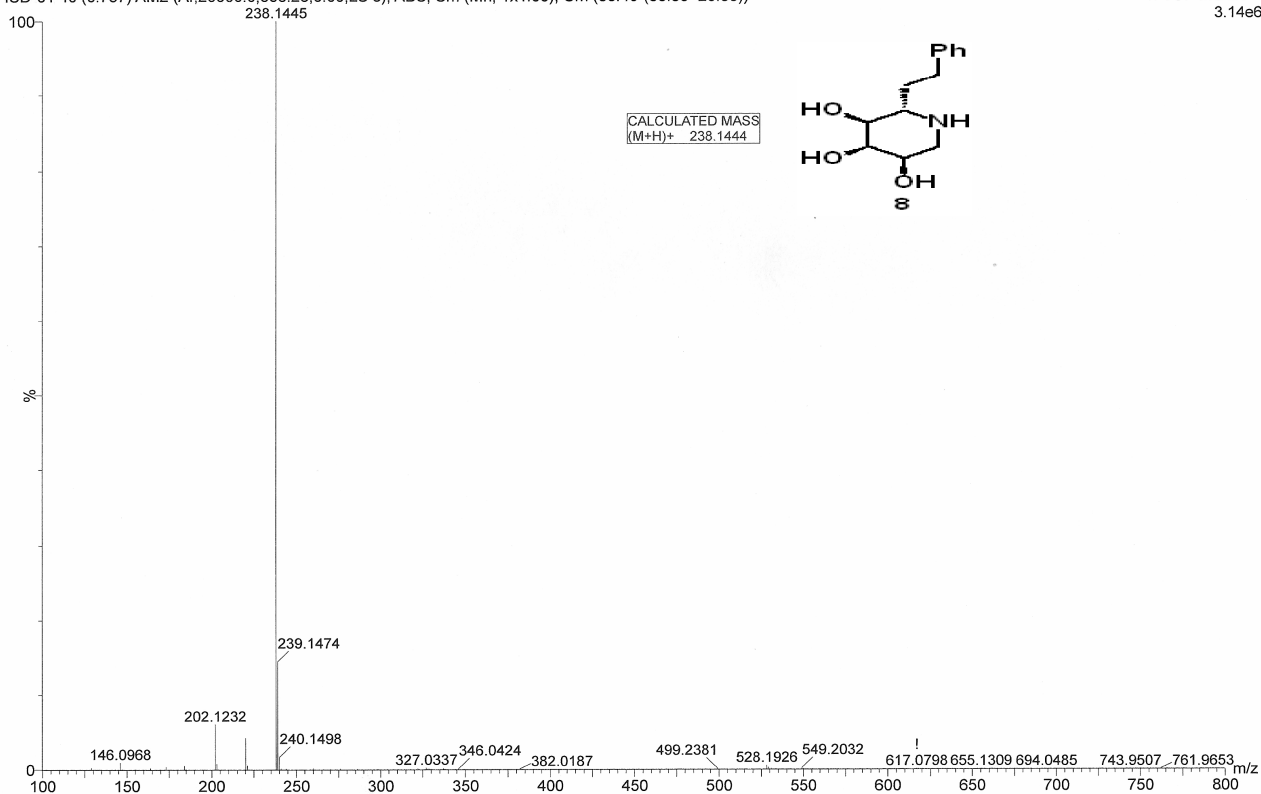


ISD-01

IISER PUNE

ISD-01 40 (0.757) AM2 (Ar,20000.0,556.28,0.00,LS 3); ABS; Sm (Mn, 1x1.00); Cm (38:40-(53:63+20:33))

1: TOF MS ES+
3.14e6



CALCULATED MASS
(M+H)+ 238.1444



Supplementary Figure 24S. Human Bgal enzyme activity inhibition curves in presence of increasing concentration [μM] of compounds 7a-7e and 8. Fraction of bGal enzyme activity remaining was calculated as bgal Activity in presence of compound/bgal activity in absence of any compound. Activity was monitored using MU-bGal (0.45 mM) at pH 4.5. Reactions were performed in triplicate. Error bars associated with each data point represents standard deviation ($n=3$). Nonlinear curve fitting to extract IC_{50} values was performed within GraphPad Prism 5.2

