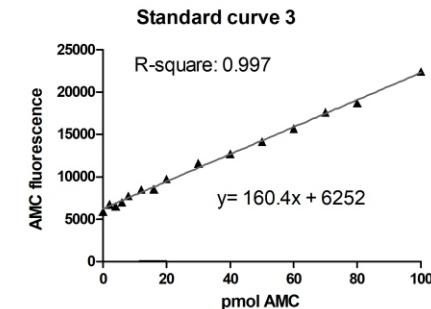
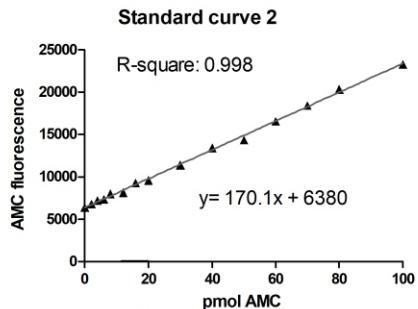
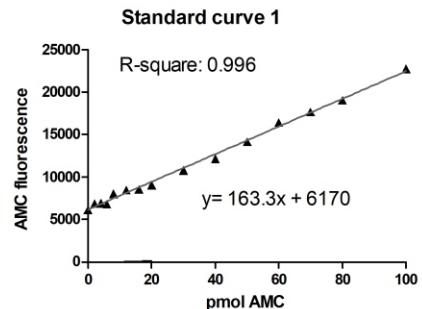
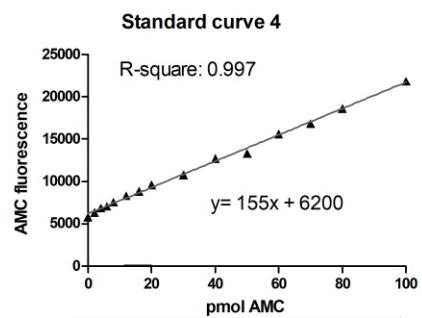


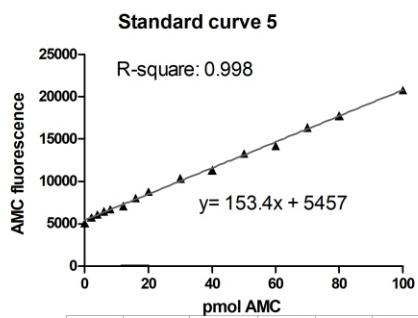
Supplementary Figure S1



	pmol AMC in serum matrix	Fluorescence	pmol AMC	% of expected value	% deviation
S1	0	6138	-0.267018		
S2	2	6891	4.3869861	219.35	-13.83
S3	4	6943	4.705594	117.64	17.64
S4	6	6814	3.9152013	65.25	-16.07
S5	8	8028	11.341217	141.77	-13.77
S6	12	8466	14.03713	116.98	-16.98
S7	16	8544	14.515042	90.72	-9.28
S8	20	9042	17.566326	87.83	-12.17
S9	30	10825	28.490901	94.97	-5.03
S10	40	12153	36.627658	91.57	-8.43
S11	50	14125	48.710251	97.42	-2.58
S12	60	16427	62.814779	104.69	4.69
S13	70	17686	70.528767	100.76	0.76
S14	80	19027	78.745175	98.43	-1.57
S15	100	22770	101.67882	101.68	1.68



	pmol AMC in serum matrix	Fluorescence	pmol AMC	% of expected value	% deviation
S1	0	5803	-3.0064935		
S2	2	6343	0.5	25.00	-19.60
S3	4	6875	3.9545455	98.86	-1.14
S4	6	7064	5.1818182	86.36	-13.64
S5	8	7582	8.5454545	106.82	6.82
S6	12	8305	13.24026	110.34	10.34
S7	16	8874	16.933065	105.84	5.84
S8	20	9599	21.642857	108.21	8.21
S9	30	10784	29.337662	97.79	-2.21
S10	40	12744	42.064935	105.16	5.16
S11	50	13292	45.623377	91.25	-8.75
S12	60	15643	60.88961	101.48	1.48
S13	70	16852	68.74026	98.20	-1.80
S14	80	18606	80.13987	100.16	0.16
S15	100	21847	101.17532	101.18	1.18



	pmol AMC in serum matrix	Fluorescence	pmol AMC	% of expected value	% deviation
S1	0	5081	-2.8815789		
S2	2	5759	1.57894737	78.95	-21.05
S3	4	6121	3.96052632	99.01	-0.99
S4	6	6501	6.46052632	107.68	7.68
S5	8	6713	7.85526316	98.19	-1.81
S6	12	7111	10.4736842	87.28	-12.72
S7	16	8025	16.4868421	103.04	3.04
S8	20	8806	21.625	108.13	8.13
S9	30	10366	31.8881579	106.29	6.29
S10	40	11327	38.2105263	95.53	-4.47
S11	50	13308	51.2434211	102.49	2.49
S12	60	14206	57.1513158	95.25	-4.75
S13	70	16366	71.3618421	101.95	1.95
S14	80	17760	80.5328947	100.67	0.67
S15	100	20771	100.342105	100.34	0.34

Supplementary Figure S1: Linearity and lower limit of quantification (LLOQ). AMC-standards were pipetted as outlined in Table 1. Raw AMC fluorescence (in artificial units) was related to the corresponding AMC amount in pmol. The slope, y-intercept and, R-square were calculated by linear regression analysis. The data for the independent experiments used for Figure 1 are shown. Concentrations of the calibration standards were back-calculated. Green numbers indicate back-calculation of the standards within the requested limits ($\pm 15\%$ of the nominal value, for LLOQ $\pm 20\%$), yellow colors refer to back calculations close, but outside the requested range, and red colors indicate values where back-calculation was inaccurate. LLOQ of each standard curve was determined to range between 2 and 6 pmol (= uppermost green AMC standard) and was averaged to obtain the mean LLOQ (i.e. 4.8 pmol AMC).

Supplementary Table S1: Coefficients of variation (CV) of the individual measurements of within-run and between-run precision.

Within-run precision (Data from Figure 3)		Between-run precision healthy donors (Data from Figure 4A)		Between-run precision Patients (Data from Figure 4B)	
Sample ID	CV (%)	Sample ID	CV (%)	Sample ID	CV (%)
Healthy donor 2	3.73	Healthy donor 1	8.21	Patient 1	9.81
Healthy donor 3	3.22	Healthy donor 2	6.98	Patient 2	48.94
Healthy donor 5	1.06	Healthy donor 3	9.25	Patient 3	17.63
Healthy donor 6	3.57	Healthy donor 4	4.93	Patient 4	105.82
Healthy donor 8	1.81	Healthy donor 5	6.54	Patient 5	52.14
Healthy donor 10	4.45	Healthy donor 6	10.09	Patient 6	26.19
Patient 1	6.83	Healthy donor 8	12.76	Patient 7	22.31
Patient 7	3.68	Healthy donor 10	7.23	Patient 8	65.24
Patient 8	5.22			Patient 9	128.58
Patient 14	5.98			Patient 10	19.74
Patient 18	5.16			Patient 11	23.66
Patient 19	14.78			Patient 12	86.78
				Patient 13	32.81
				Patient 14	66.66
				Patient 15	62.11
				Patient 16	43.14
				Patient 17	65.04
				Patient 18	37.23
				Patient 19	47.03
				Patient 20	41.37