

## 8 Přílohy

### 8.1 Hodnoty respirace dětských kontrolních linií fibroblastů po kultivaci v mediu s obsahem glukózy

Jednotka pmol/(s\*mg)

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
K1	P9	51,1586	21,2454	25,0923	76,5535	109,3589	102,1055	89,7226	32,3881	90,6592	5,1647	292,7246	N/A
K1	P9	60,8269	19,2197	28,2207	81,1234	116,2356	113,0697	93,9678	38,0142	96,6885	4,9276	308,284	N/A
K1	P10	53,35	13,7142	25,7593	76,14	109,6644	108,2103	85,7001	33,2038	90,8659	1,908	330,1578	99,8729
K1	P12	68,7069	26,2191	33,0811	100,3143	139,9674	126,8385	112,8504	42,8916	130,1953	1,4408	441,0381	150,9704
K1	P12	53,8244	8,6669	21,7761	72,9015	116,7767	111,312	94,1929	37,9221	109,4219	0,2241	344,6783	124,5926
K2	P13	71,312	23,474	38,7162	94,4182	129,94	128,8766	104,764	41,4005	112,8558	3,8002	334,5831	109,0157
K2	P14	75,1673	29,0877	49,1092	59,8522	98,8297	99,8098	56,865	28,9851	57,7523	3,3245	326,7342	104,8303
K2	P14	59,9406	22,9982	37,4062	73,4688	123,5597	120,9861	89,7637	59,5175	85,3315	3,5686	375,2255	108,732
K3	P+5	63,4321	27,0978	38,4254	64,0144	86,5559	86,0198	51,1945	18,9408	49,9144	2,9015	N/A	N/A
K3	P+5	63,0365	24,4908	26,2424	75,5356	110,5132	106,6805	88,4957	32,1746	93,2446	8,0978	379,9013	148,0795
K3	P+5	65,5494	18,4752	32,1343	93,1769	134,185	127,6719	99,1742	33,8527	117,1541	6,9523	375,0441	122,0678
K4	P+5	45,752	10,6217	20,0566	54,5026	50,5616	50,6454	34,6302	18,2036	35,6306	2,7567	426,0557	N/A
K4	P+5	53,9843	10,9138	18,431	75,5275	108,9453	104,3463	56,2288	17,1395	55,708	1,7968	320,6461	176,8955
K4	P+5	55,3242	12,8367	18,2396	71,3306	105,1894	106,2465	77,6449	25,7572	73,8422	6,0718	355,5143	182,4314
	<b>Průměr</b>	<b>60,0975</b>	<b>19,2187</b>	<b>29,4779</b>	<b>76,3471</b>	<b>110,0202</b>	<b>106,6299</b>	<b>81,0853</b>	<b>32,8851</b>	<b>85,6617</b>	<b>3,7811</b>	<b>354,6605</b>	<b>132,7488</b>
	<b>Medián</b>	<b>60,3838</b>	<b>20,2326</b>	<b>27,2316</b>	<b>75,5316</b>	<b>110,0888</b>	<b>107,4454</b>	<b>89,1092</b>	<b>32,7960</b>	<b>90,7626</b>	<b>3,4466</b>	<b>344,6783</b>	<b>123,3302</b>

## 8.2 Poměry respirace dětských kontrolních linií fibroblastů po kultivaci v mediu s obsahem glukózy

ID	Pasáž	CI	OXPPOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPPOS	CII/OXPPOS	CIV/OXPPOS	leak/OXPPOS	FCCP/OXPPOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
K1	P9	71,39	104,19	96,94	84,56	N/A	27,22	85,49	0,69	0,81	N/A	0,26	0,82
K1	P9	76,20	111,31	108,14	89,04	N/A	33,09	91,76	0,68	0,80	N/A	0,30	0,82
K1	P10	74,23	107,76	106,30	83,79	230,28	31,30	88,96	0,69	0,78	2,14	0,29	0,83
K1	P12	98,87	138,53	125,40	111,41	290,07	41,45	128,75	0,71	0,80	2,09	0,30	0,93
K1	P12	72,68	116,55	111,09	93,97	220,09	37,70	109,20	0,62	0,81	1,89	0,32	0,94
K2	P13	90,62	126,14	125,08	100,96	225,57	37,60	109,06	0,72	0,80	1,79	0,30	0,86
K2	P14	56,53	95,51	96,49	53,54	221,90	25,66	54,43	0,59	0,56	2,32	0,27	0,57
K2	P14	69,90	119,99	117,42	86,20	266,49	55,95	81,76	0,58	0,72	2,22	0,47	0,68
K3	P+5	86,22	127,23	120,72	92,22	252,98	26,90	110,20	0,68	0,72	1,99	0,21	0,87
K3	P+5	61,11	83,65	83,12	48,29	N/A	16,04	47,01	0,73	0,58	N/A	0,19	0,56
K3	P+5	67,44	102,42	98,58	80,40	231,82	24,08	85,15	0,66	0,79	2,26	0,24	0,83
K4	P+5	51,75	47,80	47,89	31,87	N/A	15,45	32,87	1,08	0,67	N/A	0,32	0,69
K4	P+5	73,73	107,15	102,55	54,43	143,75	15,34	53,91	0,69	0,51	1,34	0,14	0,50
K4	P+5	65,26	99,12	100,17	71,57	173,08	19,69	67,77	0,66	0,72	1,75	0,20	0,68
	<b>Průměr</b>	<b>72,57</b>	<b>106,24</b>	<b>102,85</b>	<b>77,30</b>	<b>225,60</b>	<b>29,10</b>	<b>81,88</b>	<b>0,70</b>	<b>0,72</b>	<b>1,98</b>	<b>0,27</b>	<b>0,76</b>
	<b>Medián</b>	<b>72,03</b>	<b>107,45</b>	<b>104,43</b>	<b>84,18</b>	<b>227,93</b>	<b>27,06</b>	<b>85,32</b>	<b>0,68</b>	<b>0,75</b>	<b>2,04</b>	<b>0,28</b>	<b>0,82</b>

### 8.3 Hodnoty respirace dětských kontrolních linií fibroblastů po kultivaci v mediu s obsahem galaktózy

Jednotka pmol/(s\*mg)

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
K1	P16	34,3081	15,2723	52,2677	100,5403	145,4981	142,3364	113,2159	63,9262	121,4644	5,6802	429,0401	121,1563
K1	P16	31,7044	17,6716	46,7607	101,9464	150,405	147,3394	121,0023	59,9427	130,5602	6,3957	421,7852	137,3773
K1	P13	23,9764	14,4695	35,3682	54,187	98,9866	95,8562	65,4738	41,4087	59,1571	3,1204	276,2897	85,5814
K1	P14	34,0891	15,5488	49,3051	76,6811	116,2441	115,8985	79,5543	40,8436	97,2278	1,5934	345,4877	59,6774
K1	P14	31,1361	15,4649	45,7952	63,2787	92,3782	91,7673	58,1668	29,4882	65,9324	2,8857	270,7306	75,2001
K2	P15	27,0249	17,9991	48,7828	55,3742	79,1824	81,4194	26,9069	18,0242	20,6099	5,8446	335,9831	89,4814
K2	P15	28,8039	22,4328	57,5249	71,3613	99,242	101,2674	37,6038	28,5061	33,138	6,3227	369,5224	89,1509
K2	P15	55,1701	37,4282	56,8926	70,9838	79,9260	77,3292	28,0526	25,1788	40,3617	5,9812	337,5310	79,3694
K4	P+5	41,8391	20,2466	37,8131	61,7763	73,2663	73,0351	29,9905	19,3812	28,438	6,1612	332,9366	133,2118
K4	P+5	44,6651	21,045	44,415	57,164	83,2635	80,9476	51,3448	27,207	51,0718	6,8751	458,3658	144,176
K4	P+5	60,2023	27,167	39,7698	60,0247	87,5568	88,3461	59,8721	26,9706	68,2935	8,1496	417,0828	127,0824
	Průměr	37,5381	20,4314	46,7905	70,3016	100,5408	99,5948	61,0167	34,6252	65,1141	5,3645	363,1595	103,7695
	Medián	34,0891	17,9991	46,7607	63,2787	92,3782	91,7673	58,1668	28,5061	59,1571	5,9812	345,4877	89,4814

#### 8.4 Poměry respirace dětských kontrolních linií fibroblastů po kultivaci v mediu s obsahem galaktózy

ID	Pasáž	CI	OXPPOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPPOS	CII/OXPPOS	CIV/OXPPOS	leak/OXPPOS	FCCP/OXPPOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>K1</b>	<b>P16</b>	94,86	139,82	136,66	107,54	307,88	58,25	115,78	0,68	0,77	2,20	0,42	0,83
<b>K1</b>	<b>P16</b>	95,55	144,01	140,94	114,61	284,41	53,55	124,16	0,66	0,80	1,97	0,37	0,86
<b>K1</b>	<b>P13</b>	51,07	95,87	92,74	62,35	190,71	38,29	56,04	0,53	0,65	1,99	0,40	0,58
<b>K1</b>	<b>P14</b>	75,09	114,65	114,31	77,96	285,81	39,25	95,63	0,65	0,68	2,49	0,34	0,83
<b>K1</b>	<b>P14</b>	60,39	89,49	88,88	55,28	195,53	26,60	63,05	0,67	0,62	2,18	0,30	0,70
<b>K2</b>	<b>P15</b>	49,53	73,34	75,57	21,06	246,50	12,18	14,77	0,68	0,29	3,36	0,17	0,20
<b>K2</b>	<b>P15</b>	65,04	92,92	94,94	31,28	280,37	22,18	26,82	0,70	0,34	3,02	0,24	0,29
<b>K2</b>	<b>P15</b>	65,00	73,94	71,35	22,07	258,16	19,20	34,38	0,88	0,30	3,49	0,26	0,46
<b>K4</b>	<b>P+5</b>	55,62	67,11	66,87	23,83	199,72	13,22	22,28	0,83	0,36	2,98	0,20	0,33
<b>K4</b>	<b>P+5</b>	50,29	76,39	74,07	44,47	314,19	20,33	44,20	0,66	0,58	4,11	0,27	0,58
<b>K4</b>	<b>P+5</b>	51,88	79,41	80,20	51,72	290,00	18,82	60,14	0,65	0,65	3,65	0,24	0,76
	<b>Průměr</b>	<b>66,67</b>	<b>95,18</b>	<b>94,23</b>	<b>58,47</b>	<b>262,04</b>	<b>30,19</b>	<b>64,43</b>	<b>0,69</b>	<b>0,55</b>	<b>2,86</b>	<b>0,29</b>	<b>0,59</b>
	<b>Medián</b>	<b>62,70</b>	<b>89,49</b>	<b>88,88</b>	<b>53,50</b>	<b>282,39</b>	<b>24,39</b>	<b>58,09</b>	<b>0,67</b>	<b>0,62</b>	<b>2,98</b>	<b>0,27</b>	<b>0,58</b>

## 8.5 Hodnoty respirace dospělých kontrolních linií fibroblastů po kultivaci v mediu s obsahem glukózy

Jednotka pmol/(s\*mg)

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
K5	P11	40,4434	13,0382	13,5566	37,1571	42,0427	35,5764	22,4365	8,2697	29,069	1,2908	295,3579	158,4532
K5	P11	63,6072	34,3613	48,5972	55,3768	79,7036	82,6979	36,2349	20,3003	35,1342	4,3027	359,4734	132,3871
K5	P11	42,8442	8,8802	16,2826	22,8825	80,7075	72,4035	26,8454	17,6259	25,754	-4,3747	358,6511	230,2013
	<b>Průměr</b>	<b>48,9649</b>	<b>18,7599</b>	<b>26,1455</b>	<b>38,4721</b>	<b>67,4846</b>	<b>63,5593</b>	<b>28,5056</b>	<b>15,3986</b>	<b>29,9857</b>	<b>0,4063</b>	<b>337,8275</b>	<b>173,6805</b>
	<b>Medián</b>	<b>42,8442</b>	<b>13,0382</b>	<b>16,2826</b>	<b>37,1571</b>	<b>79,7036</b>	<b>72,4035</b>	<b>26,8454</b>	<b>17,6259</b>	<b>29,0690</b>	<b>1,2908</b>	<b>358,6511</b>	<b>158,4532</b>

## 8.6 Poměry respirace dospělých kontrolních linií fibroblastů po kultivaci v mediu s obsahem glukózy

ID	Pasáž	CI	OXPHOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPHOS	CII/OXPHOS	CIV/OXPHOS	leak/OXPHOS	FCCP/OXPHOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
K5	P11	35,87	40,75	34,29	21,15	136,90	6,98	27,78	0,88	0,52	3,36	0,17	0,68
K5	P11	51,07	75,40	78,40	31,93	227,09	16,00	30,83	0,68	0,42	3,01	0,21	0,41
K5	P11	27,26	85,08	76,78	31,22	128,45	22,00	30,13	0,32	0,37	1,51	0,26	0,35
	<b>Průměr</b>	<b>38,07</b>	<b>67,08</b>	<b>63,15</b>	<b>28,10</b>	<b>164,15</b>	<b>14,99</b>	<b>29,58</b>	<b>0,63</b>	<b>0,44</b>	<b>2,63</b>	<b>0,21</b>	<b>0,48</b>
	<b>Medián</b>	<b>35,87</b>	<b>75,40</b>	<b>76,78</b>	<b>31,22</b>	<b>136,90</b>	<b>16,00</b>	<b>30,13</b>	<b>0,68</b>	<b>0,42</b>	<b>3,01</b>	<b>0,21</b>	<b>0,41</b>

## 8.7 Hodnoty respirace dospělých kontrolních linií fibroblastů po kultivaci v mediu s obsahem galaktózy

Jednotka pmol/(s\*mg)

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
K5	P13	39,4159	18,8782	28,9458	49,4994	86,4382	86,2924	66,9	39,9907	75,8473	4,2094	339,2473	140,5657
K5	P15	41,362	15,927	34,4735	50,4887	81,1573	83,6875	58,9291	25,9496	70,2007	6,637	395,7347	155,4268
K5	P15	36,3333	17,7641	39,1065	52,2609	71,9476	79,4063	37,972	24,9522	45,3546	4,2756	458,4052	216,1073
	<b>Průměr</b>	<b>39,0371</b>	<b>17,5231</b>	<b>34,1753</b>	<b>50,7497</b>	<b>79,8477</b>	<b>83,1287</b>	<b>54,6004</b>	<b>30,2975</b>	<b>63,8009</b>	<b>5,0407</b>	<b>397,7957</b>	<b>170,6999</b>
	<b>Medián</b>	<b>39,4159</b>	<b>17,7641</b>	<b>34,4735</b>	<b>50,4887</b>	<b>81,1573</b>	<b>83,6875</b>	<b>58,9291</b>	<b>25,9496</b>	<b>70,2007</b>	<b>4,2756</b>	<b>395,7347</b>	<b>155,4268</b>

## 8.8 Poměry respirace dospělých kontrolních linií fibroblastů po kultivaci v mediu s obsahem galaktózy

ID	Pasáž	CI	OXPHOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPHOS	CII/OXPHOS	CIV/OXPHOS	leak/OXPHOS	FCCP/OXPHOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
K5	P13	45,29	82,23	82,08	62,69	198,68	35,78	71,64	0,55	0,76	2,42	0,44	0,87
K5	P15	43,85	74,52	77,05	52,29	240,31	19,31	63,56	0,59	0,70	3,22	0,26	0,85
K5	P15	47,99	67,67	75,13	33,70	242,30	20,68	41,08	0,71	0,50	3,58	0,31	0,61
	<b>Průměr</b>	<b>45,71</b>	<b>74,81</b>	<b>78,09</b>	<b>49,56</b>	<b>227,10</b>	<b>25,26</b>	<b>58,76</b>	<b>0,62</b>	<b>0,65</b>	<b>3,07</b>	<b>0,33</b>	<b>0,78</b>
	<b>Medián</b>	<b>45,29</b>	<b>74,52</b>	<b>77,05</b>	<b>52,29</b>	<b>240,31</b>	<b>20,68</b>	<b>63,56</b>	<b>0,59</b>	<b>0,70</b>	<b>3,22</b>	<b>0,31</b>	<b>0,85</b>

## 8.9 Hodnoty respirace fibroblastů pacientů po kultivaci v mediu s obsahem glukózy

Jednotka pmol/(s\*mg)

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
P1	P+6	45,5534	23,437	34,9376	42,1963	71,6322	72,0153	51,2279	24,5706	60,3224	2,5522	330,8751	111,3955
P1	P+6	36,5536	14,6026	19,9646	32,1125	77,3146	73,1519	37,0982	22,1095	31,4193	1,9397	349,581	229,2213
P1	P+6	32,8406	6,3101	10,7008	43,0066	63,9206	61,4	56,2169	21,6782	41,2043	2,0794	374,4905	260,1918
	<b>Průměr</b>	<b>38,3159</b>	<b>14,7832</b>	<b>21,8677</b>	<b>39,1051</b>	<b>70,9558</b>	<b>68,8557</b>	<b>48,1810</b>	<b>22,7861</b>	<b>44,3153</b>	<b>2,1904</b>	<b>351,6489</b>	<b>200,2695</b>
	<b>Medián</b>	<b>36,5536</b>	<b>14,6026</b>	<b>19,9646</b>	<b>42,1963</b>	<b>71,6322</b>	<b>72,0153</b>	<b>51,2279</b>	<b>22,1095</b>	<b>41,2043</b>	<b>2,0794</b>	<b>349,5810</b>	<b>229,2213</b>

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
P2	P+5	44,4072	17,768	14,2967	40,897	54,4725	54,3542	39,2158	14,2903	40,2436	0,5191	191,8391	106,6708
P2	P+5	42,3292	7,9477	13,4665	42,3644	59,9568	52,304	40,123	12,0554	44,2277	1,497	184,1667	105,7399
P2	P+5	46,7242	16,9241	14,8659	47,2772	64,896	56,437	50,1583	14,8615	52,7295	0,0694	212,0485	98,4029
	<b>Průměr</b>	<b>44,4869</b>	<b>14,2133</b>	<b>14,2097</b>	<b>43,5129</b>	<b>59,7751</b>	<b>54,3651</b>	<b>43,1657</b>	<b>13,7357</b>	<b>45,7336</b>	<b>0,6952</b>	<b>196,0181</b>	<b>103,6045</b>
	<b>Medián</b>	<b>44,4072</b>	<b>16,9241</b>	<b>14,2967</b>	<b>42,3644</b>	<b>59,9568</b>	<b>54,3542</b>	<b>40,1230</b>	<b>14,2903</b>	<b>44,2277</b>	<b>0,5191</b>	<b>191,8391</b>	<b>105,7399</b>

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
P3	P+5	84,8815	23,1807	29,3259	79,074	112,603	107,7426	83,3823	32,2687	112,9826	5,6848	407,9359	137,7966
P3	P+5	74,0432	24,7907	26,8925	74,3655	103,2971	106,0345	88,3452	32,6405	107,7752	8,9211	438,861	162,8714
P3	P+5	59,883	22,6007	20,6409	79,3611	107,7443	104,7714	75,6477	32,2379	76,9256	10,9744	400,2031	233,9127
	<b>Průměr</b>	<b>72,9359</b>	<b>23,5240</b>	<b>25,6198</b>	<b>77,6002</b>	<b>107,8815</b>	<b>106,1828</b>	<b>82,4584</b>	<b>32,3824</b>	<b>99,2278</b>	<b>8,5268</b>	<b>415,6667</b>	<b>178,1936</b>
	<b>Medián</b>	<b>74,0432</b>	<b>23,1807</b>	<b>26,8925</b>	<b>79,0740</b>	<b>107,7443</b>	<b>106,0345</b>	<b>83,3823</b>	<b>32,2687</b>	<b>107,7752</b>	<b>8,9211</b>	<b>407,9359</b>	<b>162,8714</b>

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
P4	P+4	32,9106	10,0637	16,2684	28,0416	60,1614	45,3198	35,6401	12,2792	35,5317	1,9211	201,1253	82,4161
P4	P+4	24,8825	5,0409	13,5804	20,4652	31,6273	30,4207	27,5671	6,6694	25,4602	2,0064	289,5173	190,216
P4	P+5	39,3784	8,5445	12,7405	29,8533	56,1335	52,3877	41,7314	16,0277	39,6666	1,3798	244,8194	139,0144
P4	P+7	45,4416	17,8352	22,4808	38,5893	62,896	62,4417	44,4682	15,0418	48,7336	2,6348	232,8846	109,7632
P4	P+8	31,6472	7,8911	8,3506	31,5807	75,3786	68,6810	44,5657	13,5454	37,0230	1,6530	200,6138	109,8232
	<b>Průměr</b>	<b>35,3381</b>	<b>9,8277</b>	<b>14,2879</b>	<b>30,1220</b>	<b>56,5092</b>	<b>53,4825</b>	<b>39,5842</b>	<b>12,8222</b>	<b>37,7201</b>	<b>1,9178</b>	<b>241,9578</b>	<b>137,2034</b>
	<b>Medián</b>	<b>35,5142</b>	<b>8,2173</b>	<b>14,1520</b>	<b>13,1605</b>	<b>30,7167</b>	<b>59,5148</b>	<b>57,4147</b>	<b>43,0998</b>	<b>14,2959</b>	<b>38,3433</b>	<b>1,8282</b>	<b>238,8520</b>



ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
P5	P+5	36,9333	17,7974	27,155	46,7416	72,0545	73,4418	53,4135	22,3018	55,741	6,0682	761,5432	405,91
P5	P+5	34,822	22,9184	21,8659	32,0572	39,1668	44,2348	18,4774	13,9577	20,9934	8,8282	450,441	279,2319
P5	P+5	31,7792	9,5634	21,0581	24,625	38,0116	36,9784	22,7451	12,9735	20,6025	7,2674	477,2197	309,8915
	<b>Průměr</b>	<b>34,5115</b>	<b>16,7597</b>	<b>23,3597</b>	<b>34,4746</b>	<b>49,7443</b>	<b>51,5517</b>	<b>31,5453</b>	<b>16,4110</b>	<b>32,4456</b>	<b>7,3879</b>	<b>563,0680</b>	<b>331,6778</b>
	<b>Medián</b>	<b>34,8220</b>	<b>17,7974</b>	<b>21,8659</b>	<b>32,0572</b>	<b>39,1668</b>	<b>44,2348</b>	<b>22,7451</b>	<b>13,9577</b>	<b>20,9934</b>	<b>7,2674</b>	<b>477,2197</b>	<b>309,8915</b>

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
P6	P+5	51,7262	19,5151	28,6348	59,0084	78,0481	71,2973	53,1815	29,7429	60,6786	4,7586	332,6714	178,8385
P6	P+5	72,7152	16,1497	25,3296	70,1943	99,1819	98,4054	84,1442	31,3912	96,7293	4,1671	413,4818	187,5457
P6	P+5	56,9678	16,4042	21,5284	61,8042	81,146	66,9321	57,6449	24,8734	78,2387	4,5621	505,2266	289,7266
	<b>Průměr</b>	<b>60,4697</b>	<b>17,3563</b>	<b>25,1643</b>	<b>63,6690</b>	<b>86,1253</b>	<b>78,8783</b>	<b>64,9902</b>	<b>28,6692</b>	<b>78,5489</b>	<b>4,4959</b>	<b>417,1266</b>	<b>218,7036</b>
	<b>Medián</b>	<b>56,9678</b>	<b>16,4042</b>	<b>25,3296</b>	<b>61,8042</b>	<b>81,1460</b>	<b>71,2973</b>	<b>57,6449</b>	<b>29,7429</b>	<b>78,2387</b>	<b>4,5621</b>	<b>413,4818</b>	<b>187,5457</b>

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
P7	P+5	39,6873	11,2979	22,3475	40,7088	60,8973	60,4508	38,2183	10,5980	45,3759	1,4969	246,0496	117,5858
P7	P+5	44,9233	21,2132	22,2319	40,8786	72,0637	67,5708	31,9020	14,0897	30,2086	-0,9086	289,0268	134,6974
	<b>Průměr</b>	<b>42,3053</b>	<b>16,2556</b>	<b>22,2897</b>	<b>40,7937</b>	<b>66,4805</b>	<b>64,0108</b>	<b>35,0602</b>	<b>12,3439</b>	<b>37,7923</b>	<b>0,2942</b>	<b>267,5382</b>	<b>126,1416</b>
	<b>Medián</b>	<b>42,3053</b>	<b>16,2556</b>	<b>22,2897</b>	<b>40,7937</b>	<b>66,4805</b>	<b>64,0108</b>	<b>35,0602</b>	<b>12,3439</b>	<b>37,7923</b>	<b>0,2942</b>	<b>267,5382</b>	<b>126,1416</b>

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
P8	P+5	28,3084	14,5184	20,5977	24,2431	36,2776	38,1661	18,7049	7,601	20,5915	2,211	238,1294	125,7843
P8	P+6	54,1937	22,6619	33,6519	39,8564	65,2126	64,3646	35,2041	15,3726	40,0605	3,7581	330,7342	150,0447
P8	P+6	30,4128	13,2053	27,2214	29,4333	46,3831	45,3527	21,2578	10,1220	21,9362	3,4652	273,3020	117,3035
	<b>Průměr</b>	<b>37,6383</b>	<b>16,7952</b>	<b>27,1570</b>	<b>31,1776</b>	<b>49,2911</b>	<b>49,2945</b>	<b>25,0556</b>	<b>11,0319</b>	<b>27,5294</b>	<b>3,1448</b>	<b>280,7219</b>	<b>131,0442</b>
	<b>Medián</b>	<b>30,4128</b>	<b>14,5184</b>	<b>27,2214</b>	<b>29,4333</b>	<b>46,3831</b>	<b>45,3527</b>	<b>21,2578</b>	<b>10,1220</b>	<b>21,9362</b>	<b>3,4652</b>	<b>273,3020</b>	<b>125,7843</b>

### 8.10 Poměry respirace fibroblastů pacientů po kultivaci v mediu s obsahem glukózy

ID	Pasáž	CI	OXPPOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPPOS	CII/OXPPOS	CIV/OXPPOS	leak/OXPPOS	FCCP/OXPPOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>P1</b>	<b>P+6</b>	39,64	69,08	69,46	48,68	219,48	22,02	57,77	0,57	0,70	3,18	0,32	0,84
<b>P1</b>	<b>P+6</b>	30,17	75,37	71,21	35,16	120,36	20,17	29,48	0,40	0,47	1,60	0,27	0,39
<b>P1</b>	<b>P+6</b>	40,93	61,84	59,32	54,14	114,30	19,60	39,12	0,66	0,88	1,85	0,32	0,63
	<b>Průměr</b>	<b>36,91</b>	<b>68,77</b>	<b>66,67</b>	<b>45,99</b>	<b>151,38</b>	<b>20,60</b>	<b>42,12</b>	<b>0,55</b>	<b>0,68</b>	<b>2,21</b>	<b>0,30</b>	<b>0,62</b>
	<b>Medián</b>	<b>39,64</b>	<b>69,08</b>	<b>69,46</b>	<b>48,68</b>	<b>120,36</b>	<b>20,17</b>	<b>39,12</b>	<b>0,57</b>	<b>0,70</b>	<b>1,85</b>	<b>0,32</b>	<b>0,63</b>

ID	Pasáž	CI	OXPPOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPPOS	CII/OXPPOS	CIV/OXPPOS	leak/OXPPOS	FCCP/OXPPOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>P2</b>	<b>P+5</b>	40,38	53,95	53,84	38,70	85,17	13,77	39,72	0,75	0,72	1,58	0,26	0,74
<b>P2</b>	<b>P+5</b>	40,87	58,46	50,81	38,63	78,43	10,56	42,73	0,70	0,66	1,34	0,18	0,73
<b>P2</b>	<b>P+5</b>	47,21	64,83	56,37	50,09	113,65	14,79	52,66	0,73	0,77	1,75	0,23	0,81
	<b>Průměr</b>	<b>42,82</b>	<b>59,08</b>	<b>53,67</b>	<b>42,47</b>	<b>92,41</b>	<b>13,04</b>	<b>45,04</b>	<b>0,73</b>	<b>0,72</b>	<b>1,56</b>	<b>0,22</b>	<b>0,76</b>
	<b>Medián</b>	<b>40,87</b>	<b>58,46</b>	<b>53,84</b>	<b>38,70</b>	<b>85,17</b>	<b>13,77</b>	<b>42,73</b>	<b>0,73</b>	<b>0,72</b>	<b>1,58</b>	<b>0,23</b>	<b>0,74</b>

ID	Pasáž	CI	OXPPOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPPOS	CII/OXPPOS	CIV/OXPPOS	leak/OXPPOS	FCCP/OXPPOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>P3</b>	<b>P+5</b>	73,39	106,92	102,06	77,70	270,14	26,58	107,30	0,69	0,73	2,53	0,25	1,00
<b>P3</b>	<b>P+5</b>	65,44	94,38	97,11	79,42	275,99	23,72	98,85	0,69	0,84	2,92	0,25	1,05
<b>P3</b>	<b>P+5</b>	68,39	96,77	93,80	64,67	166,29	21,26	65,95	0,71	0,67	1,72	0,22	0,68
	<b>Průměr</b>	<b>69,07</b>	<b>99,35</b>	<b>97,66</b>	<b>73,93</b>	<b>237,47</b>	<b>23,86</b>	<b>90,70</b>	<b>0,70</b>	<b>0,75</b>	<b>2,39</b>	<b>0,24</b>	<b>0,91</b>
	<b>Medián</b>	<b>68,39</b>	<b>96,77</b>	<b>97,11</b>	<b>77,70</b>	<b>270,14</b>	<b>23,72</b>	<b>98,85</b>	<b>0,69</b>	<b>0,73</b>	<b>2,53</b>	<b>0,25</b>	<b>1,00</b>

ID	Pasáž	CI	OXPPOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPPOS	CII/OXPPOS	CIV/OXPPOS	leak/OXPPOS	FCCP/OXPPOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>P4</b>	<b>P+4</b>	26,12	58,24	43,40	33,72	118,71	10,36	33,61	0,45	0,58	2,04	0,18	0,58
<b>P4</b>	<b>P+4</b>	18,46	29,62	28,41	25,56	99,30	4,66	23,45	0,62	0,86	3,35	0,16	0,79
<b>P4</b>	<b>P+5</b>	28,47	54,75	51,01	40,35	105,81	14,65	38,29	0,52	0,74	1,93	0,27	0,70
<b>P4</b>	<b>P+7</b>	35,95	60,26	59,81	41,83	123,12	12,41	46,10	0,60	0,69	2,04	0,21	0,76
<b>P4</b>	<b>P+8</b>	29,93	73,73	67,03	42,92	90,79	11,90	35,37	0,41	0,58	1,23	0,16	0,48
	<b>Průměr</b>	<b>27,79</b>	<b>55,32</b>	<b>49,93</b>	<b>36,88</b>	<b>107,55</b>	<b>10,80</b>	<b>35,36</b>	<b>0,52</b>	<b>0,69</b>	<b>2,12</b>	<b>0,19</b>	<b>0,66</b>
	<b>Medián</b>	<b>28,47</b>	<b>58,24</b>	<b>51,01</b>	<b>40,35</b>	<b>105,81</b>	<b>11,90</b>	<b>35,37</b>	<b>0,52</b>	<b>0,69</b>	<b>2,04</b>	<b>0,18</b>	<b>0,70</b>

ID	Pasáž	CI	OXPPOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPPOS	CII/OXPPOS	CIV/OXPPOS	leak/OXPPOS	FCCP/OXPPOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>P5</b>	<b>P+5</b>	40,67	65,99	67,37	47,35	355,63	16,23	49,67	0,62	0,72	5,39	0,25	0,75
<b>P5</b>	<b>P+5</b>	23,23	30,34	35,41	9,65	171,21	5,13	12,17	0,77	0,32	5,64	0,17	0,40
<b>P5</b>	<b>P+5</b>	17,36	30,74	29,71	15,48	167,33	5,71	13,34	0,56	0,50	5,44	0,19	0,43
	<b>Průměr</b>	<b>27,09</b>	<b>42,36</b>	<b>44,16</b>	<b>24,16</b>	<b>231,39</b>	<b>9,02</b>	<b>25,06</b>	<b>0,65</b>	<b>0,51</b>	<b>5,49</b>	<b>0,20</b>	<b>0,53</b>
	<b>Medián</b>	<b>23,23</b>	<b>30,74</b>	<b>35,41</b>	<b>15,48</b>	<b>171,21</b>	<b>5,71</b>	<b>13,34</b>	<b>0,62</b>	<b>0,50</b>	<b>5,44</b>	<b>0,19</b>	<b>0,43</b>

ID	Pasáž	CI	OXPPOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPPOS	CII/OXPPOS	CIV/OXPPOS	leak/OXPPOS	FCCP/OXPPOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>P6</b>	<b>P+5</b>	54,25	73,29	66,54	48,42	153,83	24,98	55,92	0,74	0,66	2,10	0,34	0,76
<b>P6</b>	<b>P+5</b>	66,03	95,01	94,24	79,98	225,94	27,22	92,56	0,69	0,84	2,38	0,29	0,97
<b>P6</b>	<b>P+5</b>	57,24	76,58	62,37	53,08	215,50	20,31	73,68	0,75	0,69	2,81	0,27	0,96
	<b>Průměr</b>	<b>59,17</b>	<b>81,63</b>	<b>74,38</b>	<b>60,49</b>	<b>198,42</b>	<b>24,17</b>	<b>74,05</b>	<b>0,73</b>	<b>0,73</b>	<b>2,43</b>	<b>0,30</b>	<b>0,90</b>
	<b>Medián</b>	<b>57,24</b>	<b>76,58</b>	<b>66,54</b>	<b>53,08</b>	<b>215,50</b>	<b>24,98</b>	<b>73,68</b>	<b>0,74</b>	<b>0,69</b>	<b>2,38</b>	<b>0,29</b>	<b>0,96</b>

ID	Pasáž	CI	OXPPOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPPOS	CII/OXPPOS	CIV/OXPPOS	leak/OXPPOS	FCCP/OXPPOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>P7</b>	<b>P+5</b>	39,21	59,40	58,95	36,72	128,46	9,10	43,88	0,66	0,62	2,16	0,15	0,74
<b>P7</b>	<b>P+5</b>	41,79	72,97	68,48	32,81	154,33	15,00	31,12	0,57	0,45	2,11	0,21	0,43
	<b>Průměr</b>	<b>40,50</b>	<b>66,19</b>	<b>63,72</b>	<b>34,77</b>	<b>141,40</b>	<b>12,05</b>	<b>37,50</b>	<b>0,62</b>	<b>0,53</b>	<b>2,14</b>	<b>0,18</b>	<b>0,58</b>
	<b>Medián</b>	<b>57,24</b>	<b>76,58</b>	<b>66,54</b>	<b>53,08</b>	<b>215,50</b>	<b>12,05</b>	<b>37,50</b>	<b>0,62</b>	<b>0,53</b>	<b>2,14</b>	<b>0,18</b>	<b>0,58</b>

ID	Pasáž	CI	OXPPOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPPOS	CII/OXPPOS	CIV/OXPPOS	leak/OXPPOS	FCCP/OXPPOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>P8</b>	<b>P+5</b>	22,03	34,07	35,96	16,49	112,35	5,39	18,38	0,65	0,48	3,30	0,16	0,54
<b>P8</b>	<b>P+6</b>	36,10	61,45	60,61	31,45	180,69	11,61	36,30	0,59	0,51	2,94	0,19	0,59
<b>P8</b>	<b>P+6</b>	25,97	42,92	41,89	17,79	156,00	6,66	18,47	0,61	0,41	3,63	0,16	0,43
	<b>Průměr</b>	<b>28,03</b>	<b>46,15</b>	<b>46,15</b>	<b>21,91</b>	<b>149,68</b>	<b>7,89</b>	<b>24,38</b>	<b>0,61</b>	<b>0,47</b>	<b>3,29</b>	<b>0,17</b>	<b>0,52</b>
	<b>Medián</b>	<b>25,97</b>	<b>42,92</b>	<b>41,89</b>	<b>17,79</b>	<b>156,00</b>	<b>6,66</b>	<b>18,47</b>	<b>0,61</b>	<b>0,48</b>	<b>3,30</b>	<b>0,16</b>	<b>0,54</b>

## 8.11 Hodnoty respirace fibroblastů pacientů po kultivaci v mediu s obsahem galaktózy

Jednotka pmol/(s\*mg)

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
P1	P+6	48,8763	21,1284	34,3945	59,8509	77,5185	79,9202	37,2308	18,4000	35,9195	4,5277	251,1886	78,1518
P1	P+6	55,8026	22,4392	35,1279	57,5958	80,3604	74,9684	44,7831	25,0382	49,0022	0,7989	272,8071	71,9564
P1	P+6	58,2655	25,0676	32,8437	49,7171	68,3621	67,6130	44,6911	21,9754	31,2926	3,4023	232,2561	69,7136
	<b>Průměr</b>	<b>54,3148</b>	<b>22,8784</b>	<b>34,1220</b>	<b>55,7213</b>	<b>75,4137</b>	<b>74,1672</b>	<b>42,2350</b>	<b>21,8045</b>	<b>38,7381</b>	<b>2,9096</b>	<b>252,0839</b>	<b>73,2739</b>
	<b>Medián</b>	<b>55,8026</b>	<b>22,4392</b>	<b>34,3945</b>	<b>57,5958</b>	<b>77,5185</b>	<b>74,9684</b>	<b>44,6911</b>	<b>21,9754</b>	<b>35,9195</b>	<b>3,4023</b>	<b>251,1886</b>	<b>71,9564</b>

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
P2	P+5	41,9471	19,0425	35,4737	58,9132	82,1481	86,2048	51,7431	24,9956	53,2456	4,7696	313,5406	129,1553
P2	P+5	30,5626	16,909	34,0064	50,4891	71,1681	69,6421	37,7945	21,6327	31,415	4,9628	235,2243	92,805
P2	P+5	44,7217	12,2477	33,3197	54,2175	76,3567	76,9466	49,3709	25,0745	54,0996	1,3376	306,344	133,9113
	<b>Průměr</b>	<b>39,0771</b>	<b>16,0664</b>	<b>34,2666</b>	<b>54,5399</b>	<b>76,5576</b>	<b>77,5978</b>	<b>46,3028</b>	<b>23,9009</b>	<b>46,2534</b>	<b>3,6900</b>	<b>285,0363</b>	<b>118,6239</b>
	<b>Medián</b>	<b>41,9471</b>	<b>16,9090</b>	<b>34,0064</b>	<b>54,2175</b>	<b>76,3567</b>	<b>76,9466</b>	<b>49,3709</b>	<b>24,9956</b>	<b>53,2456</b>	<b>4,7696</b>	<b>306,3440</b>	<b>129,1553</b>

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
P3	P+5	28,6877	15,7129	30,5687	41,5037	60,3545	68,4402	34,7695	21,1442	33,3341	5,2864	433,3207	234,595
P3	P+5	43,5652	27,4625	47,7386	60,6464	74,7472	76,2475	37,9057	27,9894	42,0502	8,9318	420,5961	161,3642
P3	P+5	53,3551	32,6885	48,0532	64,9492	83,3404	83,6174	39,887	26,3845	41,6149	5,1846	420,1479	151,8408
	<b>Průměr</b>	<b>41,8693</b>	<b>25,2880</b>	<b>42,1202</b>	<b>55,6998</b>	<b>72,8140</b>	<b>76,1017</b>	<b>37,5207</b>	<b>25,1727</b>	<b>38,9997</b>	<b>6,4676</b>	<b>424,6882</b>	<b>182,6000</b>
	<b>Medián</b>	<b>43,5652</b>	<b>27,4625</b>	<b>47,7386</b>	<b>60,6464</b>	<b>74,7472</b>	<b>76,2475</b>	<b>37,9057</b>	<b>26,3845</b>	<b>41,6149</b>	<b>5,2864</b>	<b>420,5961</b>	<b>161,3642</b>

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
P4	P+5	47,0482	11,7029	24,4906	35,221	81,8831	73,2885	53,8259	28,1482	56,6562	3,5185	306,6606	126,3233
P4	P+5	41,6269	18,6299	32,2856	42,347	65,5389	65,5942	32,1865	15,8687	31,2659	2,5104	N/A	N/A
P4	P+5	40,9152	18,9875	34,8913	52,3492	82,2707	83,3232	43,1256	24,3946	40,1899	4,3214	390,0422	171,285
	<b>Průměr</b>	<b>43,1968</b>	<b>16,4401</b>	<b>30,5558</b>	<b>43,3057</b>	<b>76,5642</b>	<b>74,0686</b>	<b>43,0460</b>	<b>22,8038</b>	<b>42,7040</b>	<b>3,4501</b>	<b>348,3514</b>	<b>148,8042</b>
	<b>Medián</b>	<b>41,6269</b>	<b>18,6299</b>	<b>32,2856</b>	<b>42,3470</b>	<b>81,8831</b>	<b>73,2885</b>	<b>43,1256</b>	<b>24,3946</b>	<b>40,1899</b>	<b>3,5185</b>	<b>348,3514</b>	<b>148,8042</b>

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
P5	P+5	52,9607	27,919	38,3205	51,49	66,3807	68,047	40,0029	24,5079	39,6382	9,2736	429,5049	186,8778
P5	P+5	36,1109	17,9258	31,2443	55,6028	84,1629	76,8501	58,9301	N/A	54,653	17,6783	593,6963	353,3854
P5	P+5	33,4618	18,6668	28,242	56,4137	81,2318	76,0606	51,2184	N/A	45,262	11,9433	643,3847	390,4656
	<b>Průměr</b>	<b>40,8445</b>	<b>21,5039</b>	<b>32,6023</b>	<b>54,5022</b>	<b>77,2585</b>	<b>73,6526</b>	<b>50,0505</b>	<b>24,5079</b>	<b>46,5177</b>	<b>12,9651</b>	<b>555,5286</b>	<b>310,2429</b>
	<b>Medián</b>	<b>36,1109</b>	<b>18,6668</b>	<b>31,2443</b>	<b>55,6028</b>	<b>81,2318</b>	<b>76,0606</b>	<b>51,2184</b>	<b>24,5079</b>	<b>45,2620</b>	<b>11,9433</b>	<b>593,6963</b>	<b>353,3854</b>



ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
P7	P+5	26,8074	19,0509	36,5685	44,2952	61,4037	66,0090	29,2977	23,2504	28,1514	3,5293	326,3870	114,5796
P7	P+5	21,4948	15,0087	34,8184	41,5725	61,8753	67,9199	35,8793	21,7150	40,6003	4,8041	280,1899	99,8166
P7	P+5	14,6652	15,2764	41,4488	47,0925	71,5860	71,4120	47,4908	23,5791	56,3182	2,6498	298,5505	98,6409
	<b>Průměr</b>	<b>20,9891</b>	<b>16,4453</b>	<b>37,6119</b>	<b>44,3201</b>	<b>64,9550</b>	<b>68,4470</b>	<b>37,5559</b>	<b>22,8482</b>	<b>41,6900</b>	<b>3,6611</b>	<b>301,7091</b>	<b>104,3457</b>
	<b>Medián</b>	<b>21,4948</b>	<b>15,2764</b>	<b>36,5685</b>	<b>44,2952</b>	<b>61,8753</b>	<b>67,9199</b>	<b>35,8793</b>	<b>23,2504</b>	<b>40,6003</b>	<b>3,5293</b>	<b>298,5505</b>	<b>99,8166</b>

ID	Pasáž	Buňky	DG	P+M+G	ADP	Suc	Cyt c	Rot	Omy	FCCP	AmA	A+T	NaN <sub>3</sub>
P8	P+5	46,304	24,9857	34,9722	38,5932	58,3628	55,468	20,0354	14,857	32,0784	-0,6042	334,6705	165,7608
P8	P+5	49,4519	30,2497	35,9282	43,9225	57,9742	59,0437	33,0070	12,1422	21,1035	3,3050	359,2449	180,7846
P8	P+5	56,6911	34,0826	44,8525	50,5887	76,6246	72,0871	41,0864	22,7650	45,1276	-0,3470	357,9187	140,8933
	<b>Průměr</b>	<b>50,8157</b>	<b>29,7727</b>	<b>38,5843</b>	<b>44,3681</b>	<b>64,3205</b>	<b>62,1996</b>	<b>31,3763</b>	<b>16,5881</b>	<b>32,7698</b>	<b>0,7846</b>	<b>350,6114</b>	<b>162,4796</b>
	<b>Medián</b>	<b>49,4519</b>	<b>30,2497</b>	<b>35,9282</b>	<b>43,9225</b>	<b>58,3628</b>	<b>59,0437</b>	<b>33,0070</b>	<b>14,8570</b>	<b>32,0784</b>	<b>-0,3470</b>	<b>357,9187</b>	<b>165,7608</b>

## 8.12 Poměry respirace fibroblastů pacientů po kultivaci v mediu s obsahem galaktózy

ID	Pasáž	CI	OXPPOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPPOS	CII/OXPPOS	CIV/OXPPOS	leak/OXPPOS	FCCP/OXPPOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>P1</b>	<b>P+6</b>	55,32	72,99	75,39	32,70	173,04	13,87	31,39	0,76	0,45	2,37	0,19	0,43
<b>P1</b>	<b>P+6</b>	56,80	79,56	74,17	43,98	200,85	24,24	48,20	0,71	0,55	2,52	0,30	0,61
<b>P1</b>	<b>P+6</b>	46,31	64,96	64,21	41,29	162,54	18,57	27,89	0,71	0,64	2,50	0,29	0,43
	<b>Průměr</b>	<b>52,81</b>	<b>72,50</b>	<b>71,26</b>	<b>39,33</b>	<b>178,81</b>	<b>18,89</b>	<b>35,83</b>	<b>0,73</b>	<b>0,55</b>	<b>2,47</b>	<b>0,26</b>	<b>0,49</b>
	<b>Medián</b>	<b>55,32</b>	<b>72,99</b>	<b>74,17</b>	<b>41,29</b>	<b>173,04</b>	<b>18,57</b>	<b>31,39</b>	<b>0,71</b>	<b>0,55</b>	<b>2,50</b>	<b>0,29</b>	<b>0,43</b>

ID	Pasáž	CI	OXPPOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPPOS	CII/OXPPOS	CIV/OXPPOS	leak/OXPPOS	FCCP/OXPPOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>P2</b>	<b>P+5</b>	54,14	77,38	81,44	46,97	184,39	20,23	48,48	0,70	0,61	2,38	0,26	0,63
<b>P2</b>	<b>P+5</b>	45,53	66,21	64,68	32,83	142,42	16,67	26,45	0,69	0,50	2,15	0,25	0,40
<b>P2</b>	<b>P+5</b>	52,88	75,02	75,61	48,03	172,43	23,74	52,76	0,70	0,64	2,30	0,32	0,70
	<b>Průměr</b>	<b>50,85</b>	<b>72,87</b>	<b>73,91</b>	<b>42,61</b>	<b>166,41</b>	<b>20,21</b>	<b>42,56</b>	<b>0,70</b>	<b>0,58</b>	<b>2,28</b>	<b>0,28</b>	<b>0,58</b>
	<b>Medián</b>	<b>52,88</b>	<b>75,02</b>	<b>75,61</b>	<b>46,97</b>	<b>172,43</b>	<b>20,23</b>	<b>48,48</b>	<b>0,70</b>	<b>0,61</b>	<b>2,30</b>	<b>0,26</b>	<b>0,63</b>

ID	Pasáž	CI	OXPHOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPHOS	CII/OXPHOS	CIV/OXPHOS	leak/OXPHOS	FCCP/OXPHOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>P3</b>	<b>P+5</b>	36,22	55,07	63,15	29,48	198,73	15,86	28,05	0,66	0,54	3,61	0,29	0,51
<b>P3</b>	<b>P+5</b>	51,71	65,82	67,32	28,97	259,23	19,06	33,12	0,79	0,44	3,94	0,29	0,50
<b>P3</b>	<b>P+5</b>	59,76	78,16	78,43	34,70	268,31	21,20	36,43	0,76	0,44	3,43	0,27	0,47
	<b>Průměr</b>	<b>49,23</b>	<b>66,35</b>	<b>69,63</b>	<b>31,05</b>	<b>242,09</b>	<b>18,71</b>	<b>32,53</b>	<b>0,74</b>	<b>0,47</b>	<b>3,66</b>	<b>0,28</b>	<b>0,49</b>
	<b>Medián</b>	<b>51,71</b>	<b>65,82</b>	<b>67,32</b>	<b>29,48</b>	<b>259,23</b>	<b>19,06</b>	<b>33,12</b>	<b>0,76</b>	<b>0,44</b>	<b>3,61</b>	<b>0,29</b>	<b>0,50</b>

ID	Pasáž	CI	OXPHOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPHOS	CII/OXPHOS	CIV/OXPHOS	leak/OXPHOS	FCCP/OXPHOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>P4</b>	<b>P+5</b>	31,70	78,36	69,77	50,31	180,34	24,63	53,14	0,40	0,64	2,30	0,31	0,68
<b>P4</b>	<b>P+5</b>	39,84	63,03	63,08	29,68	N/A	13,36	28,76	0,63	0,47		0,21	0,46
<b>P4</b>	<b>P+5</b>	48,03	77,95	79,00	38,80	218,76	20,07	35,87	0,62	0,50	2,81	0,26	0,46
	<b>Průměr</b>	<b>39,86</b>	<b>73,11</b>	<b>70,62</b>	<b>39,60</b>	<b>199,55</b>	<b>19,35</b>	<b>39,25</b>	<b>0,55</b>	<b>0,54</b>	<b>2,55</b>	<b>0,26</b>	<b>0,53</b>
	<b>Medián</b>	<b>39,84</b>	<b>77,95</b>	<b>69,77</b>	<b>38,80</b>	<b>199,55</b>	<b>20,07</b>	<b>35,87</b>	<b>0,62</b>	<b>0,50</b>	<b>2,55</b>	<b>0,26</b>	<b>0,46</b>

ID	Pasáž	CI	OXPHOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPHOS	CII/OXPHOS	CIV/OXPHOS	leak/OXPHOS	FCCP/OXPHOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>P5</b>	<b>P+5</b>	42,22	57,11	58,77	30,73	242,63	15,23	30,36	0,74	0,54	4,25	0,27	0,53
<b>P5</b>	<b>P+5</b>	37,92	66,48	59,17	41,25	240,31	N/A	36,97	0,57	0,62	3,61	N/A	0,56
<b>P5</b>	<b>P+5</b>	44,47	69,29	64,12	39,28	252,92	N/A	33,32	0,64	0,57	3,65	N/A	0,48
	<b>Průměr</b>	<b>41,54</b>	<b>64,29</b>	<b>60,69</b>	<b>37,09</b>	<b>245,29</b>	<b>15,23</b>	<b>33,55</b>	<b>0,65</b>	<b>0,58</b>	<b>3,84</b>	<b>0,27</b>	<b>0,52</b>
	<b>Medián</b>	<b>42,22</b>	<b>66,48</b>	<b>59,17</b>	<b>39,28</b>	<b>242,63</b>	<b>15,23</b>	<b>33,32</b>	<b>0,64</b>	<b>0,57</b>	<b>3,65</b>	<b>0,27</b>	<b>0,53</b>

ID	Pasáž	CI	OXPHOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPHOS	CII/OXPHOS	CIV/OXPHOS	leak/OXPHOS	FCCP/OXPHOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>P7</b>	<b>P+5</b>	40,77	57,87	62,48	25,77	211,81	19,72	24,62	0,70	0,45	3,66	0,34	0,43
<b>P7</b>	<b>P+5</b>	36,77	57,07	63,12	31,08	180,37	16,91	35,80	0,64	0,54	3,16	0,30	0,63
<b>P7</b>	<b>P+5</b>	44,44	68,94	68,76	44,84	199,91	20,93	53,67	0,64	0,65	2,90	0,30	0,78
	<b>Průměr</b>	<b>40,66</b>	<b>61,29</b>	<b>64,79</b>	<b>33,89</b>	<b>197,36</b>	<b>19,19</b>	<b>38,03</b>	<b>0,66</b>	<b>0,55</b>	<b>3,24</b>	<b>0,31</b>	<b>0,61</b>
	<b>Medián</b>	<b>40,77</b>	<b>57,87</b>	<b>63,12</b>	<b>31,08</b>	<b>199,91</b>	<b>19,72</b>	<b>35,80</b>	<b>0,64</b>	<b>0,54</b>	<b>3,16</b>	<b>0,30</b>	<b>0,63</b>

ID	Pasáž	CI	OXPPOS	Cyt c	CII	CIV	leak	FCCP	CI/OXPPOS	CII/OXPPOS	CIV/OXPPOS	leak/OXPPOS	FCCP/OXPPOS
		ADP- AmA	Suc- AmA	Cyt c- AmA	Rot- AmA	A+T- NaN <sub>3</sub>	Omy- AmA	FCCP- AmA					
<b>P8</b>	<b>P+5</b>	39,20	58,97	56,07	20,64	168,91	15,46	32,68	0,66	0,35	2,86	0,26	0,55
<b>P8</b>	<b>P+5</b>	40,62	54,67	55,74	29,70	178,46	8,84	17,80	0,74	0,54	3,26	0,16	0,33
<b>P8</b>	<b>P+5</b>	50,94	76,97	72,43	41,43	217,03	23,11	45,47	0,66	0,54	2,82	0,30	0,59
	<b>Průměr</b>	<b>43,58</b>	<b>63,54</b>	<b>61,42</b>	<b>30,59</b>	<b>188,13</b>	<b>15,80</b>	<b>31,99</b>	<b>0,69</b>	<b>0,48</b>	<b>2,98</b>	<b>0,24</b>	<b>0,49</b>
	<b>Medián</b>	<b>40,62</b>	<b>58,97</b>	<b>56,07</b>	<b>29,70</b>	<b>178,46</b>	<b>15,46</b>	<b>32,68</b>	<b>0,66</b>	<b>0,54</b>	<b>2,86</b>	<b>0,26</b>	<b>0,55</b>