

**Supplementary Table 2.** Baseline characteristics of patients with metastatic FH-deficient RCC.

<b>Characteristics</b>	<b>N (%)</b>
No. of patients	78
Age (yr), median (range)	38 (20-71)
Male, n (%)	52 (67)
Tumor size (cm), median (range)	7.5 (1.9-20.0)
Synchronous	43 (55)
Metachronous	35 (45)
ISUP grade	
2	1 (1)
3	55 (71)
4	13 (5)
NA	9 (12)
Metastatic sites	
Lymph node	48 (61)
Bone	25 (32)
Liver	10 (13)
Lung	6 (8)
Other	37 (47)
IMDC risk	
Favorable	7 (9)
Intermediate	49 (63)
Poor	13 (17)
NA	9 (12)
Treatment line	
First-line treatment	68 (87)
Second-line treatment	31 (40)
Third-line treatment	9 (12)
Fourth-line treatment	3 (4)
First-line treatment strategy	
ICI+TKI	44 (65)
TKI alone	18 (26)
Chemotherapy	2 (3)
TKI+mTORi	2 (3)
Bevacizumab+Erlotinib	1 (1)
Chemotherapy+ICI	1 (1)
Nephrectomy	69 (88)

ISUP, International Society of Urological Pathology; IMDC, International Metastatic RCC Database Consortium; NA, not available; ICI, immune check point; TKI, tyrosine kinase inhibitor; mTORi, mTOR inhibitor.