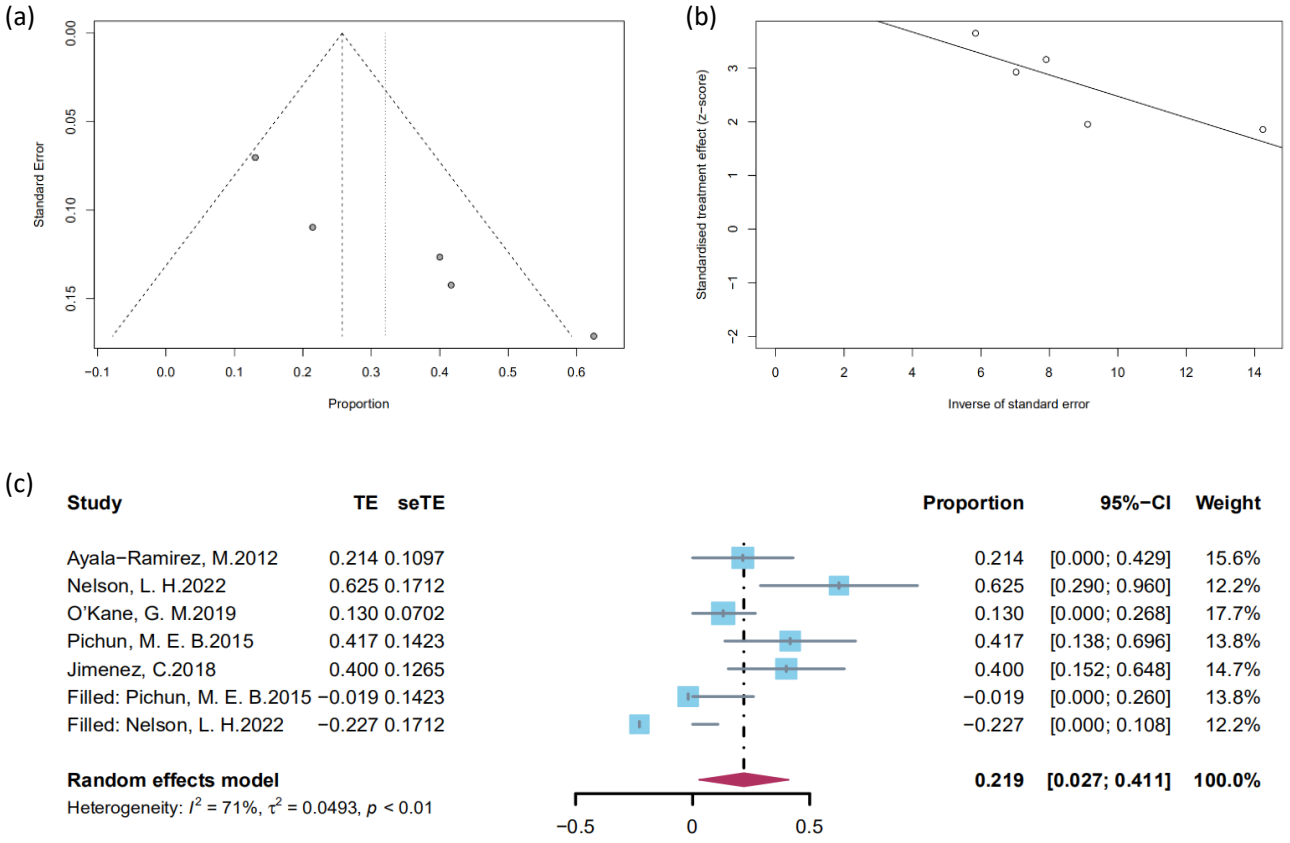
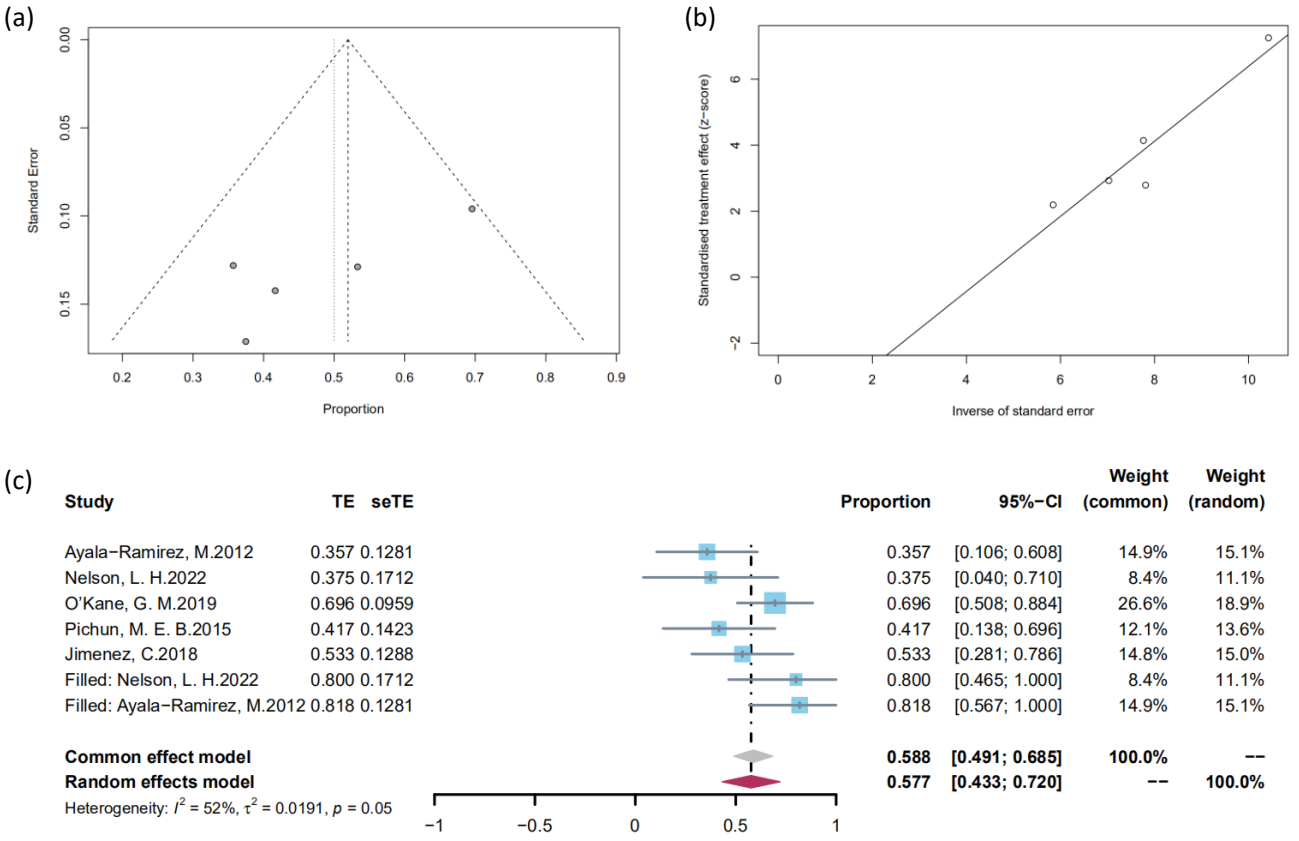
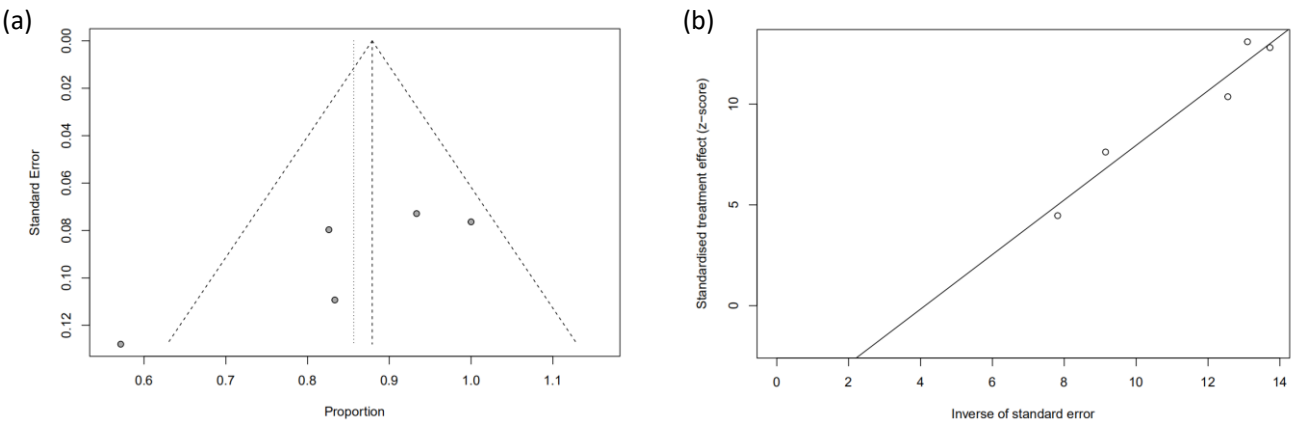
The Supplemental Materials——Figures and Tables



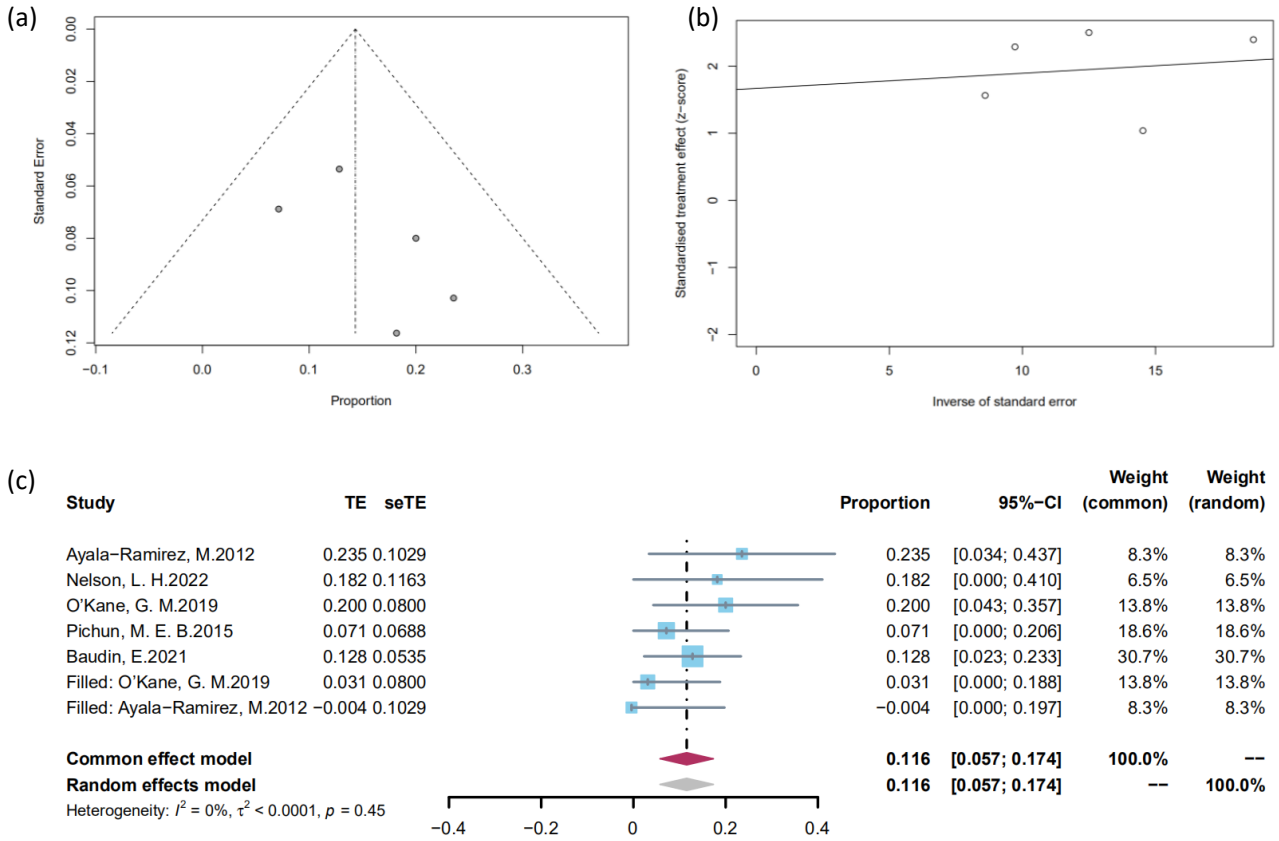
The Supplemental Figure 1. (a) The funnel plot of assessment of publication bias in five available studies with patients achieving partial response (PR). (b) Plot of Egger's test assessing the symmetry of the funnel plot. (c) Forest plot of the pooled percentage of PR after using the trill and fill method.



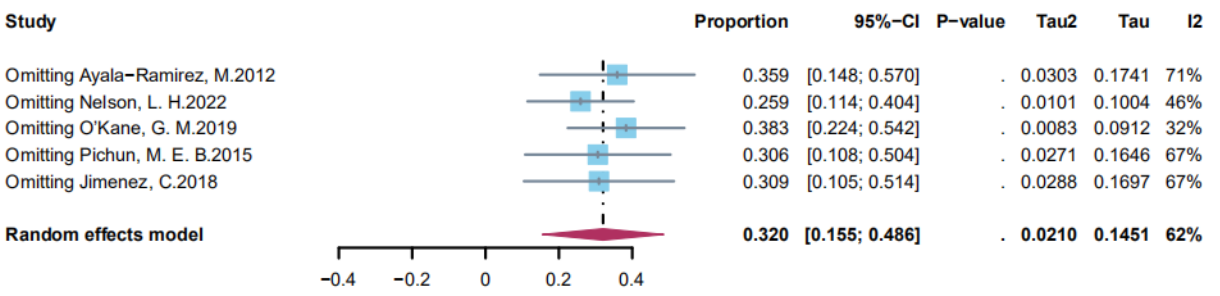
The Supplemental Figure 2. (a) The funnel plot of assessment of publication bias in five available studies with patients achieving stable disease (SD). (b) Plot of Egger's test assessing the symmetry of the funnel plot. (c) Forest plot of the pooled percentage of SD after using the trill and fill method.



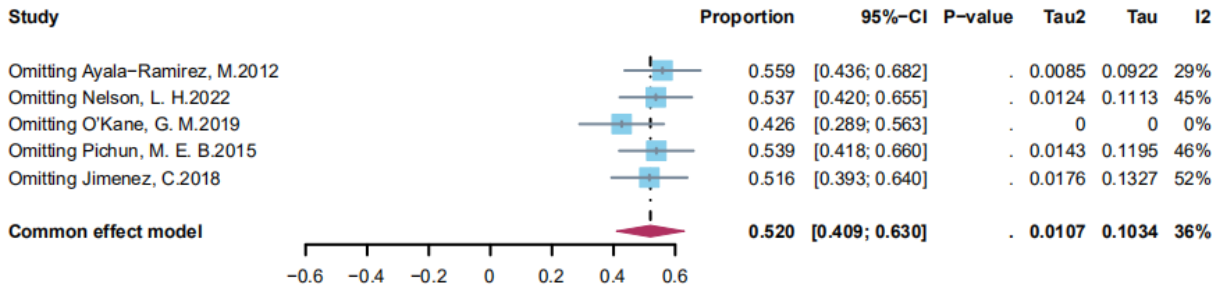
The Supplemental Figure 3. (a) The funnel plot of assessment of publication bias in five available studies with patients achieving disease control. (b) Plot of Egger's test assessing the symmetry of the funnel plot. There were no filled studies in the pooled percentage of disease control rate (DCR) after the trill and fill method.



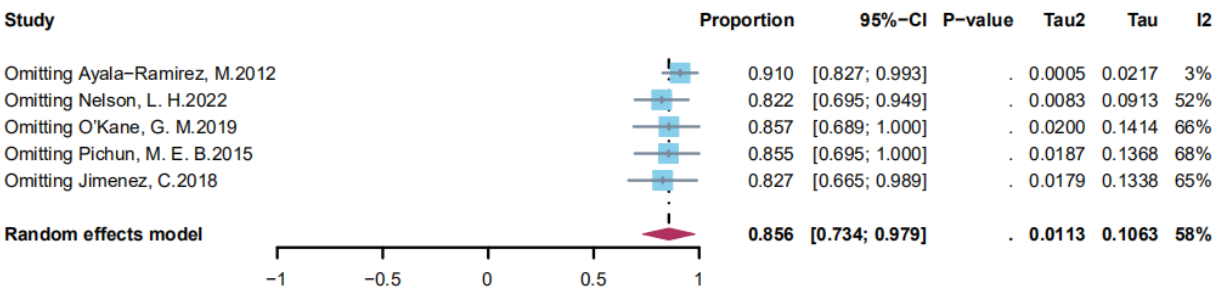
The Supplemental Figure 4. (a) The funnel plot of assessment of publication bias in five available studies with the number of discontinuation due to adverse events (AEs). (b) Plot of Egger's test assessing the symmetry of the funnel plot. (c) Forest plot of the pooled percentage of discontinuation due to AEs after using the trill and fill method.



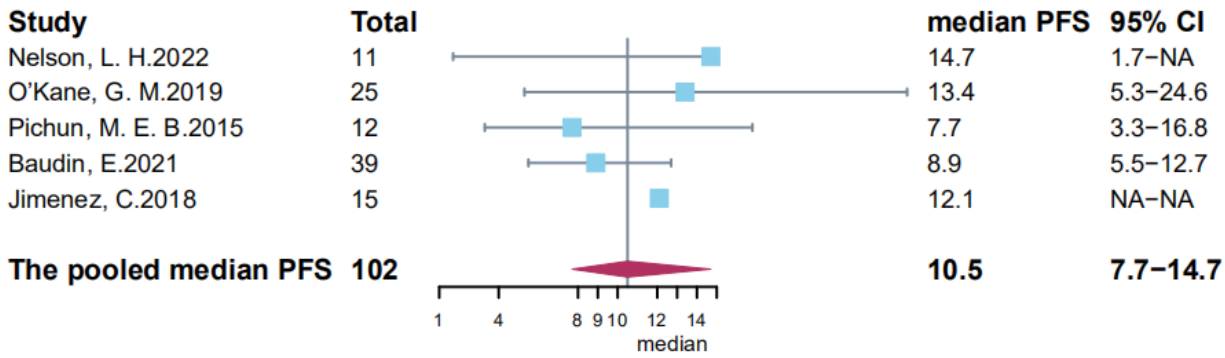
The Supplemental Figure 5. Forest plot of the sensitivity analysis of the pooled proportion of partial response (PR) through recalculating the pooled effects after omitting each study.



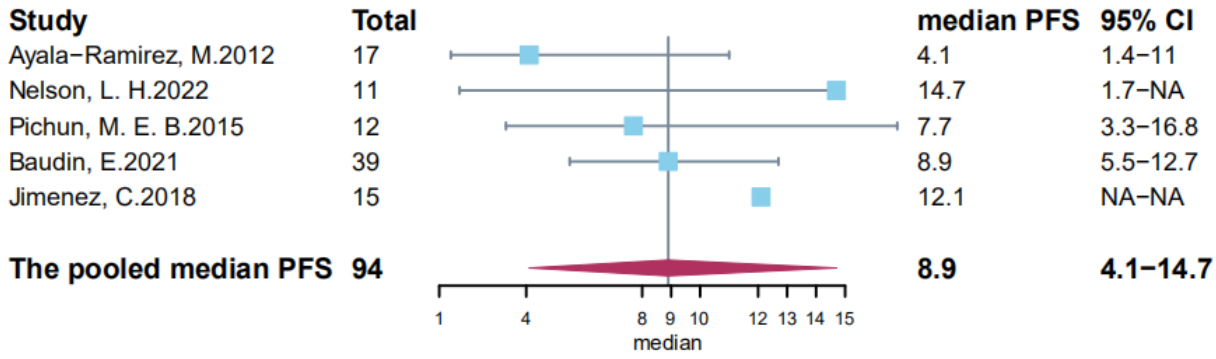
The Supplemental Figure 6. Forest plot of the sensitivity analysis of the pooled proportion of stable disease (SD) through recalculating the pooled effects after omitting each study.



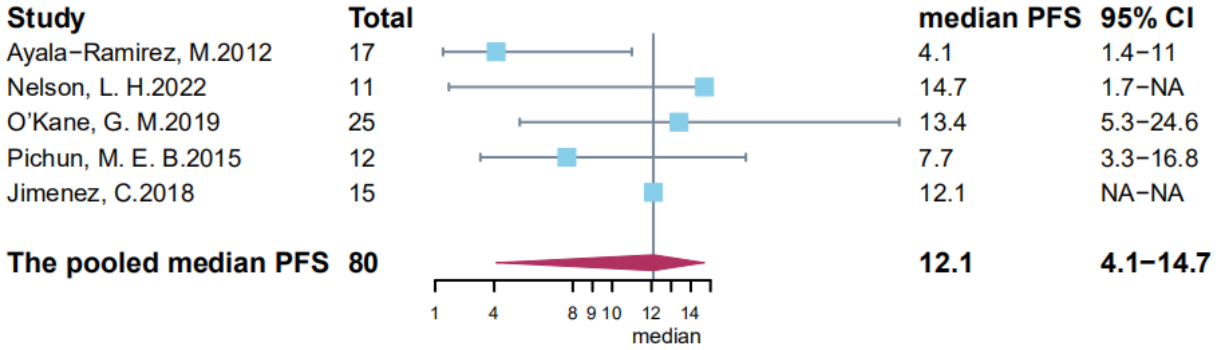
The Supplemental Figure 7. Forest plot of the sensitivity analysis of the pooled disease control rate (DCR) through recalculating the pooled effects after omitting each study.



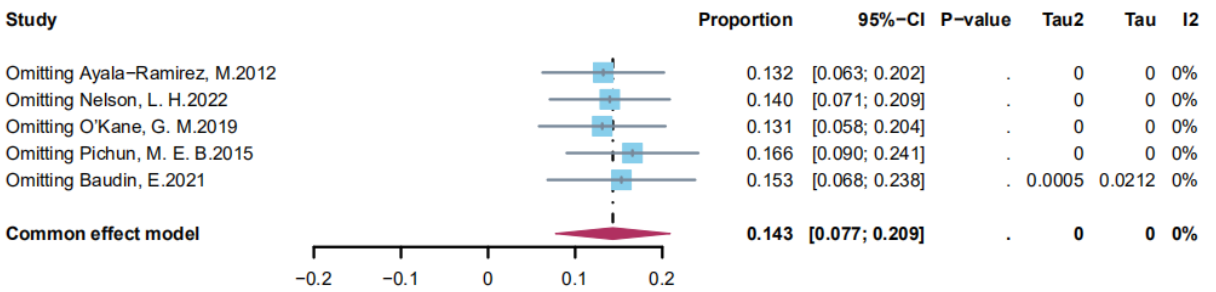
The Supplemental Figure 8. Forest plot of the sensitivity analysis of the pooled median progression-free survival (PFS) through recalculating the pooled median PFS after omitting the study of Ayala-Ramirez, M. (2012).



The Supplemental Figure 9. Forest plot of the sensitivity analysis of the pooled median progression-free survival (PFS) through recalculating the pooled median PFS after omitting the study of O'Kane, G. M. (2019).



The Supplemental Figure 10. Forest plot of the sensitivity analysis of the pooled median progression-free survival (PFS) through recalculating the pooled median PFS after omitting the study of Baudin, E.(2021).



The Supplemental Figure 11. Forest plot of the sensitivity analysis of the pooled proportion of discontinuation of drugs due to adverse events (AEs) through recalculating the pooled effects after omitting each study.

The Supplemental Table 1. Fisher's exact test of tumor response classified as partial response (PR), stable disease (SD) and progression disease (PD) in patients with pheochromocytomas (Pheo) and paragangliomas (Pgl) in the study of Ayala-Ramirez, M. (2012). Pheo, pheochromocytomas; Spgl, sympathetic paragangliomas.

Total(n=14)	Pheo(n=10)	Spgl(n=4)	Test	p
PR(n=3)	2	1	Fisher's exact test	0.091
SD(n=5)	2	3		
PD(n=6)	6	0		

The Supplemental Table 2. Fisher's exact test of tumor response classified as Response and Progression in patients with pheochromocytomas (Pheo) and paragangliomas (Pgl) in the study of Ayala-Ramirez, M. (2012). Pheo, pheochromocytomas; Spgl, sympathetic paragangliomas.

Total(n=14)	Pheo(n=10)	Spgl(n=4)	Test	p
Response(n=8)	4	4	Fisher's exact test	0.085
Progression(n=6)	6	0		

The Supplemental Table 3. Fisher's exact test of tumor response classified as partial response (PR), stable disease (SD) and progression disease (PD) in patients with *SDHB* germline mutations and non-*SDHB* mutations or undefined genetic background in the study of Ayala-Ramirez, M. (2012).

Total(n=14)	*SDHB*(n=6)	Non-*SDHB*(n=8)	Test	p
PR(n=3)	1	2	Fisher's exact test	0.149
SD(n=5)	4	1		
PD(n=6)	1	5		

The Supplemental Table 4. Fisher's exact test of tumor response classified as Response and Progression in patients with *SDHB* germline mutations and non-*SDHB* mutations or undefined genetic background in the study of Ayala-Ramirez, M. (2012).

Total(n=14)	*SDHB*(n=6)	Non-*SDHB*(n=8)	Test	p
Response(n=8)	5	3	Fisher's exact test	0.138
Progression(n=6)	1	5		

The Supplemental Table 5. Fisher's exact test of tumor response classified as partial response (PR), stable disease (SD) and progression disease (PD) in patients with *SDHB* germline mutations and non-*SDHB* mutations or undefined genetic background in the study of O'Kane, G. M. (2019).

Total(n=23)	*SDHB*(n=5)	Non-*SDHB*(n=18)	Test	p
PR(n=3)	1	2		
SD(n=16)	4	12	Fisher's exact test	0.584
PD(n=4)	0	4		

The Supplemental Table 6. Fisher's exact test of tumor response classified as Response and Progression in patients with *SDHB* germline mutations and non-*SDHB* mutations or undefined genetic background in the study of O'Kane, G. M. (2019).

Total(n=15)	*SDHB*(n=5)	Non-*SDHB*(n=18)	Test	p
Response(n=19)	5	14	Fisher's exact test	0.539
Progression(n=4)	0	4		

The Supplemental Table 7. Fisher's exact test of tumor response classified as Response and Progression in patients with *SDHB* germline mutations and non-*SDHB* mutations or undefined genetic background in the study of O'Kane, G. M. (2019).

Total(n=15)	*SDHB*(n=5)	Non-*SDHB*(n=10)	Test	P
Response(n=14)	5	14	Fisher's exact test	1
Progression(n=1)	0	1		

The Supplemental Table 8. Fisher's exact test of tumor response classified as partial response (PR), stable disease (SD) and progression disease (PD) in patients of Cluster 1and Others (no mutations related to Cluster 1 or undefined genetic background) in the study of Ayala-Ramirez, M. (2012).

Total(n=14)	Cluster 1(n=7)	Others(n=7)	Test	p
PR(n=3)	1	2		
SD(n=5)	5	0	Fisher's exact test	0.024
PD(n=6)	1	5		

The Supplemental Table 9. Fisher's exact test of tumor response classified as Response and Progression in patients of Cluster 1and Others (no mutations related to Cluster 1 or undefined genetic background) in the study of Ayala-Ramirez, M. (2012).

Total(n=14)	Cluster 1(n=7)	Other(n=7)	Test	p
Response(n=8)	6	2	Fisher's exact test	0.103
Progression(n=6)	1	5		

The Supplemental Table 10. Fisher's exact test of tumor response classified as partial response (PR), stable disease (SD) and progression disease (PD) in patients of Cluster 1and Others (no mutations related to Cluster 1 or undefined genetic background) in the study of O'Kane, G. M.(2019).

Total(n=23)	Cluster 1(n=7)	Other(n=16)	Test	p
PR(n=3)	2	1	Fisher's exact test	0.228
SD(n=16)	5	11		
PD(n=4)	0	4		

The Supplemental Table 11. Fisher's exact test of tumor response classified as Response and Progression in patients of Cluster 1and Others (no mutations related to Cluster 1 or undefined genetic background) in the study of O'Kane, G. M.(2019).

Total(n=23)	Cluster 1(n=7)	Others(n=16)	Test	P
Response(n=19)	7	12	Fisher's exact test	0.273
Progression(n=4)	0	4		