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Acute lysis of a giant pediatric adrenal cortical carcinoma following one dose of op'DDD

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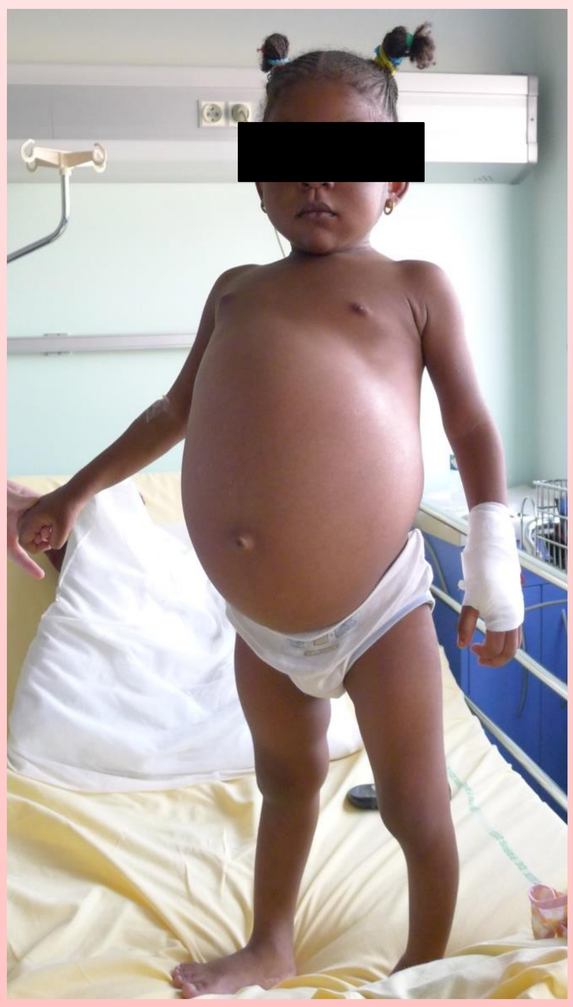
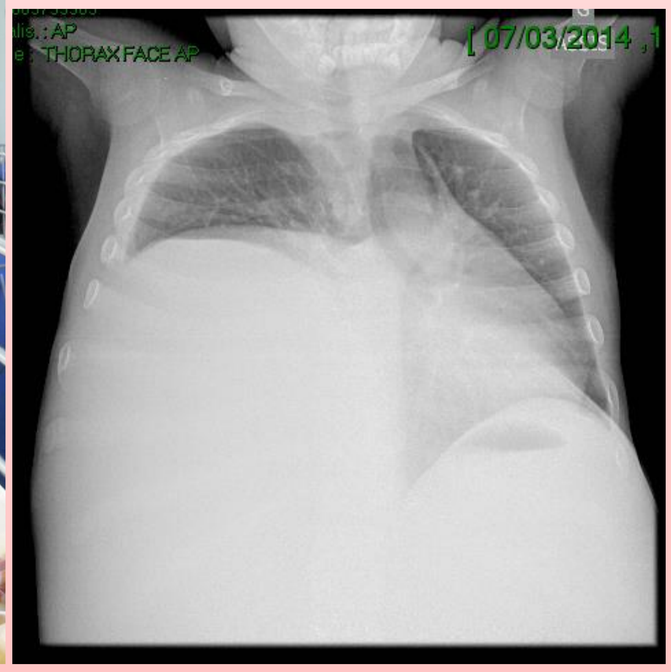
no conflict of interest

Op'DDD can be used in adrenal cortical carcinoma (ACC)
when surgery is impossible
or as an adjunct to surgery when local extension or metastases are present
We report the unexpected spectacular effects of op'DDD in an unoperable ACC





A 3-year-old black african girl presented in poor shape with a 2-year history of pubic hair, clitoromegaly, abdominal distension.
She exhibited arterial hypertension (170/120mmHg), with no sign of cardiac deficiency

The CT scan revealed a giant ACC (28 cmx25 cm) that compressed the right kidney, inferior vena cava, diaphragm, right lung, right hepatic lobe, with no metastases (panel A).

Serum testosterone was 27 ng/ml, SDHA 4270 ng/ml, FLU 155 µg/24h, midnight plamatic cortisol 22,8 µg/dl, aldosterone 18 pg/ml, renine 153 pg/ml, no elevated urinary catecholamines.

Given the risk of a primary surgery, surgeons refused to attempt tumorectomy, thus op'DDD treatment was attempted. Twelve hours after a first 500 mg dose of op'DDD, the child experienced acute intense abdominal pain and oliguria due to a typical lysis tumor syndrome with hyperkaliemia (9.4 mmol/l), hyperphosphatemia (2.8 mmol/l), hyperuricemia (572 mmol/l) and high LDL (843 UI/l).

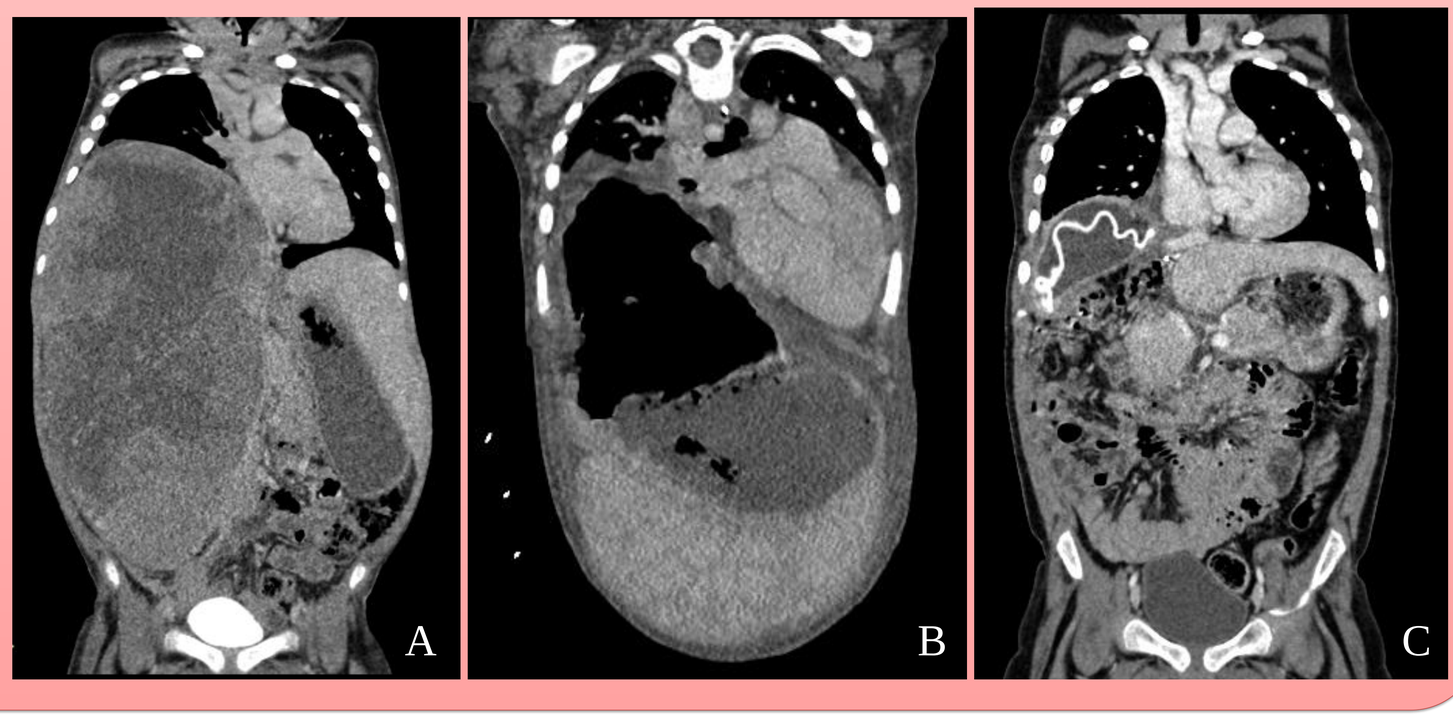
The patient went into intensive care for hemofiltration and recovered. op'DDD treatment was maintained under cover of hyperhydration and uricolytic drugs.

One week after the acute lysis, the CT scan showed that the tumor had shrinked while large zones of necrobiosis had appeared (panel B).

Six weeks later, a 14-h surgical intervention (Pr H. Martelli and Dr S. Branchereau) allowed the complete exercise of a 1.5 kg necrotic tumor, with right hepatectomy and refection of right diaphragmatic coupola .

One month later, the CT scan showed no tumoral remnants (panel C).

	Admission	48h after acute lysis syndrome	1 week after surgery	1 month after surgery
SDHEA (ng/ml)	47200	268	710	540
Testosterone (ng/ml)	27	0.65	0.47	0,21
17OHP (ng/ml)	17.4	0.22		0,15
Estradiol (pg/ml)	58	47		5
Estrone (pg/ml)	899	46	40	23



Conclusion: This case is the first to report the induction of an acute lysis of ACC by op'DDD. The lysis occured after a single dose of mitotane, and fastly decreased the hormonal secretions of the tumor. Interestingly, surgeons were able to perform a curative surgical resection a few weeks later.

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