Anesthesia in patients of aromatic L-amino acid decarboxylase (AADC) deficiency for MRI scan and stereotactic surgery

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Background

Aromatic l-amino acid decarboxylase (AADC) deficiency is a rare autosomal recessive disorder. The AADC enzyme plays an important role in the synthesis of monoamine neurotransmitter like dopamine and serotonin. Lack of the AADC enzyme would lead to reduced concentration of biogenic amines and metabolites in cerebrospinal fluid and result in autonomic dysfunction and movement disorder with symptoms such as oculogyric crises, limb dystonia, neurologic dysfunction and impaired movements. Hemodynamic instability, poor thermoregulation, and hypoglycemia due to AADC may seriously influence anesthetic managements.

Material & Methods

Inclusion criteria:

1. Definite diagnosis of AADC deficiency with the CSF study
2. Older than 24 months, or skull bones suitable for stereotactic surgery

Exclusion criteria:

1. Patient condition not suitable for surgery or anesthesia

Results

Table 1. patients' demographic data

patient	age	sex	BH	BW	MRI duration (min)	MRI ICU stay	OP duration (min)	OP ICU stay
1	6y2m	F	97	11.0	105	1	488	1
2	7y8m	M	124	20.2	75	1	689	2
3	8y	F	126	20.5	125	0.5	470	6
4	2y5m	M	84	10.1	95	3	472	3
5	2y8m	M	88	10.0	90	0	518	1
6	6y7m	F	120	17.0	70	1	524	3
7	2y7m	M	95	10.0	95	1	475	1
8	2y10m	F	91	11.0	90	1	530	2

Table 2. Anesthetic medication during lumbar puncture and MRI scan

patient	Induction			Maintenance		Extubation		
	citosol	atropine	Cistracurium	Cistracurium	Ephedrine	Atropine	Neostigmine	aminophylline
1	50	0.2	2	0	0	0	0	0
2	75	0.2	3	2	0	0	0	0
3	100	0.5	4	0	0	0.4	1	75
4	38	0.1	3	0	4	0	0	0
5	25	0.2	2	0	0	0.2	0.5	0
6	75	0.2	3	1	0	0.4	0.8	0
7	37.5	0.3	2	0	0	0.2	0.5	0
8	25	0.3	1	0	0	0.3	0.5	0

Table 3. Anesthetic medication during stereotatic surgery

patient	Induction					Maintenance						
	Citosol	Ketamine	Atropine	Cistracurium	Fentanyl	Cistracurium	Fentanyl	Norepinephrine	Dopamine	Ephedrine	Atropine	D50W
1	0	20	0.2	2	0	16	0	2mcg x7 + infusion	0	0	0	0
2	100	0	0.2	6	50	28	0	3+2+2.5 mcg	0	0	0.3	0
3	150	0	0.2	4	0	16	50	infusion	0	0	0.2	20ml
4	100	0	0.2	4	10	12	15	0	0	0	0	0
5	50	0	0	3	10	12	10	0	0	0	0	0
6	87.5	0	0.25	3	0	11	25	0	0	4	0	0
7	50	0	0.2	3	15	12	35	0	0	0	0.1!	0
8	100	0	0.2	5	0	14	0	2.5mcg	0	0	0	0

Table 1.

BH: body height, BW: body weight, OP:stereotactic surgery

Table 2.

All the units of doses are milligram(mg). Patient 1,2,4 were transferred to the ICU with endotracheal tube. Patient 4 was under severe bronchitis.

Table 3.

All the units of doses without mentioned are milligram(mg). All the patients were transferred to ICU with endotracheal tube.

Discussion and Conclusion

Due to reduced concentration of catecholamine and intact parasympathetic activity, anesthetic management in AADC deficiency patients is challenging. In our case series, hemodynamics of AADC deficiency patients were stable during general anesthesia for minor procedure such as MRI scan. In major surgeries with long surgical duration, inotropic or vasoactive support might be necessary. Our study showed that general anesthesia can be conducted safely in AADC deficiency patients with careful monitoring.