



An uncommon case of Hypertension- Multi disciplinary management

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History

- 14 year male
- Complaints: Headache, Vomiting and Giddiness. H/o of seizures.
- No voiding complaints, no h/o haematuria/ fever / pain abdomen / loss of weight or appetite.



Past history

- Hypertension diagnosed at age of 2 years when evaluated for failure to thrive;
 - Incomplete evaluation & was on Alpha Methyldopa; Not on regular follow up.
 - Birth history : Term, NVD, birth wt 2.5 kgs
 - Developmental history – appropriate to age.
 - Immunization : As per schedule
 - Family history – First born to a non consanguineous marriage. No family history of Hypertension.
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Course in Referring Hospital

- BP was 150/100; Started on T. Metoprolol, T. Alpha methyldopa, T. Thiazide and T. Nimodipine
- BUN, Creatinine was normal.
- USG Abdomen – Small left kidney
- Renal Doppler – Left Renal Artery stenosis
- Tc-DTPA - Small sized left kidney with reduced cortical function (13%) and no excretion.

USG

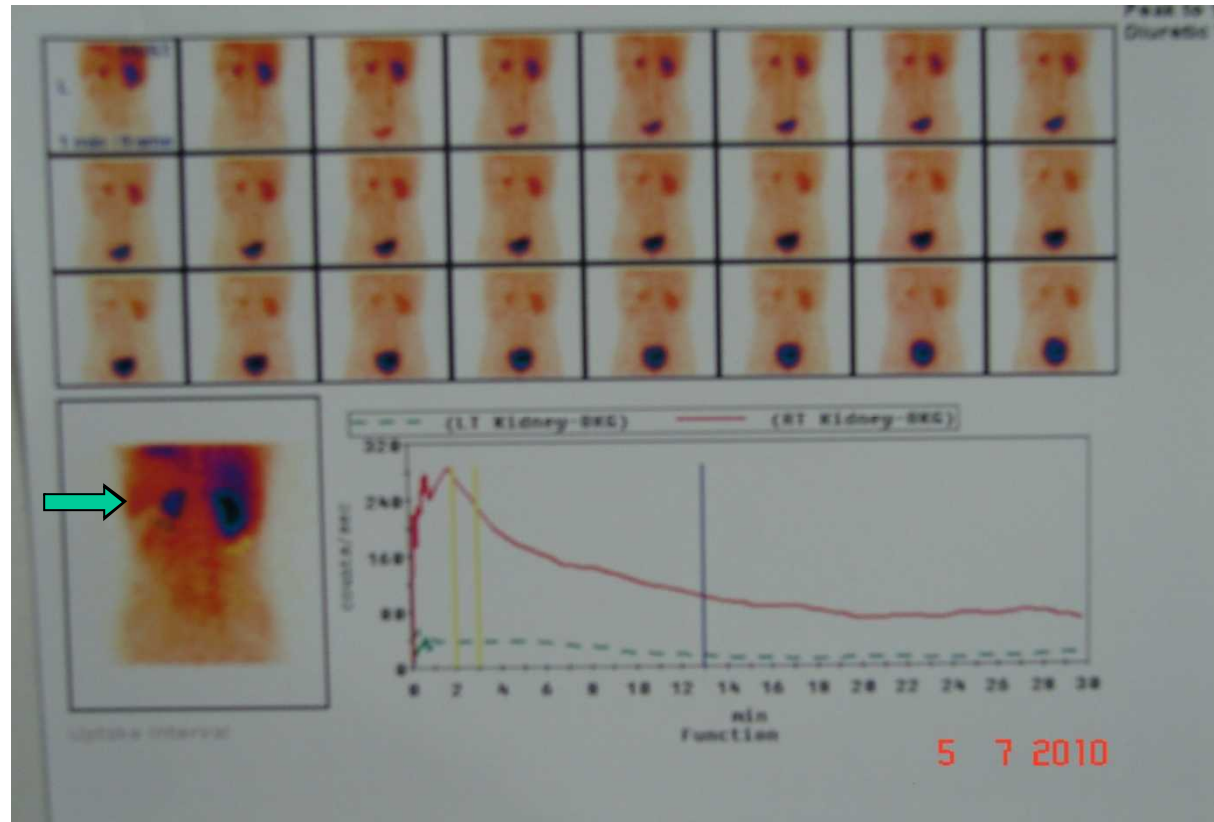
Small left kidney; normal right kidney



CT ABDOMEN



DTPA scan



Small sized left kidney with reduced cortical function (13%) and no excretion.



Course in Referring Hospital

- After 2 days, had 1 episode of generalized seizure;
- CT scan Head showed small Sub-Arachnoid Haemorrhage
- CT Angiography showed Berry Aneurysm of left Middle cerebral Artery
- Phenytoin loading given
- Referred to SRMC for further Management

CT SCAN



CT ANGIOGRAPHY





In SRMC

- Weight : 32 kg , Height : 150 cm
- HR : 84 / min
- BP : 160/110 mmHg
- Afebrile
- RR : 24 / min
- No pallor / icterus / cyanosis / clubbing / lymphadenopathy / pedal oedema



Systemic examination

- CVS : No added murmurs
- RS : Normal vesicular breath sounds
- ABDOMEN: Normal, no bruit
- CNS : No focal neurological defects.
- GCS : 15/15
- Fundus Normal



Laboratory investigations:

- Hb : 9.5 g/dl
- TC : 16160
- Poly : 87.2, Lymph : 9.2
- Platelet count : 3.31 lakhs/cu mm
- BUN : 29 , Creatinine : 0.7
- Na : 136, K : 3.0 , Cl : 85 , HCO₃ : 25
- Urine R/E :Normal
- ECHO mild LVH



Initial Management

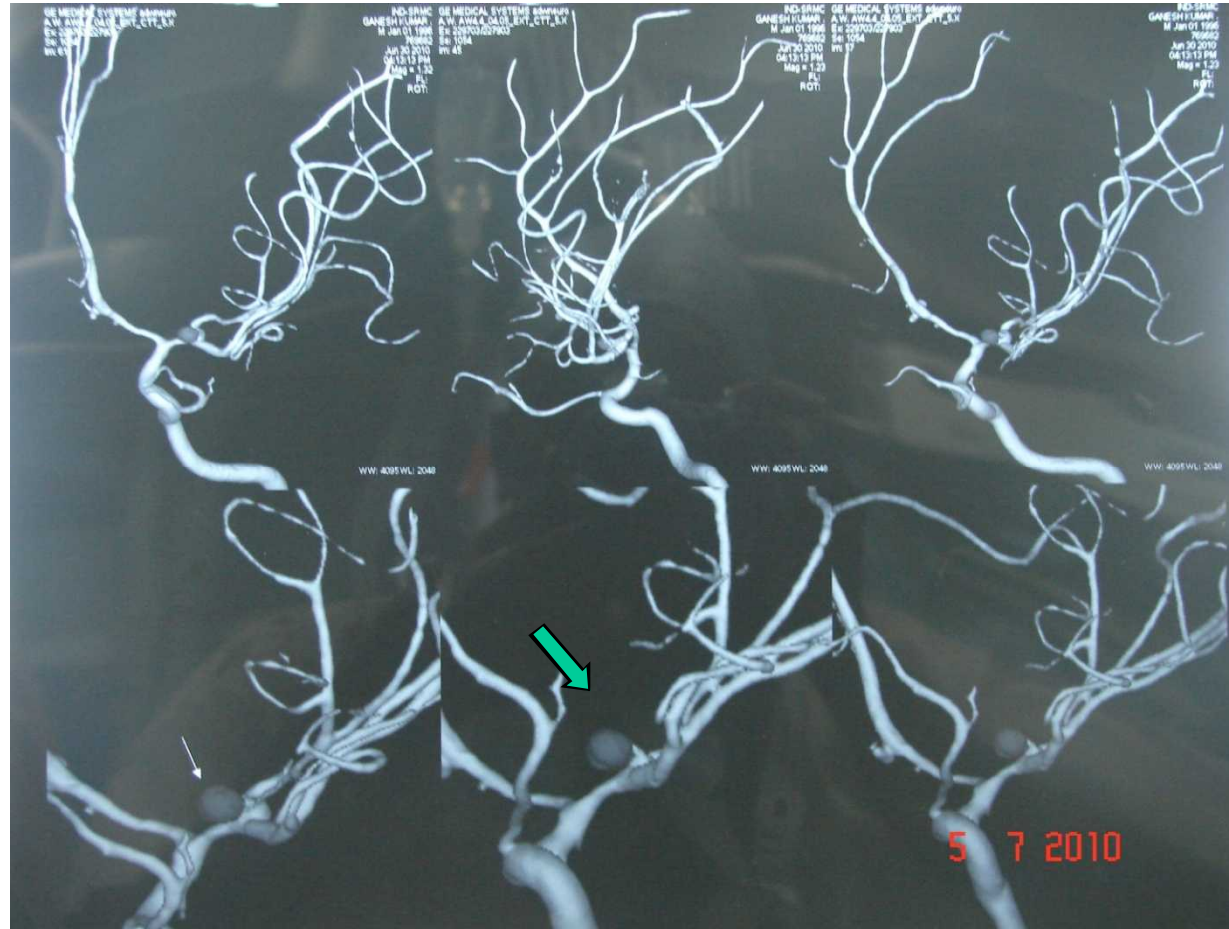
- Admitted in PICU
 - Hypertension was controlled with 4 drugs
– Metoprolol, Alpha Methyldopa, Enalapril, Nimodipine (for SAH)
 - Nephrology, Neurosurgery, Interventional radiology, Neurology & Urology Consults
 - Decided to treat aneurysm first, after Conventional Angiography, followed by nephrectomy
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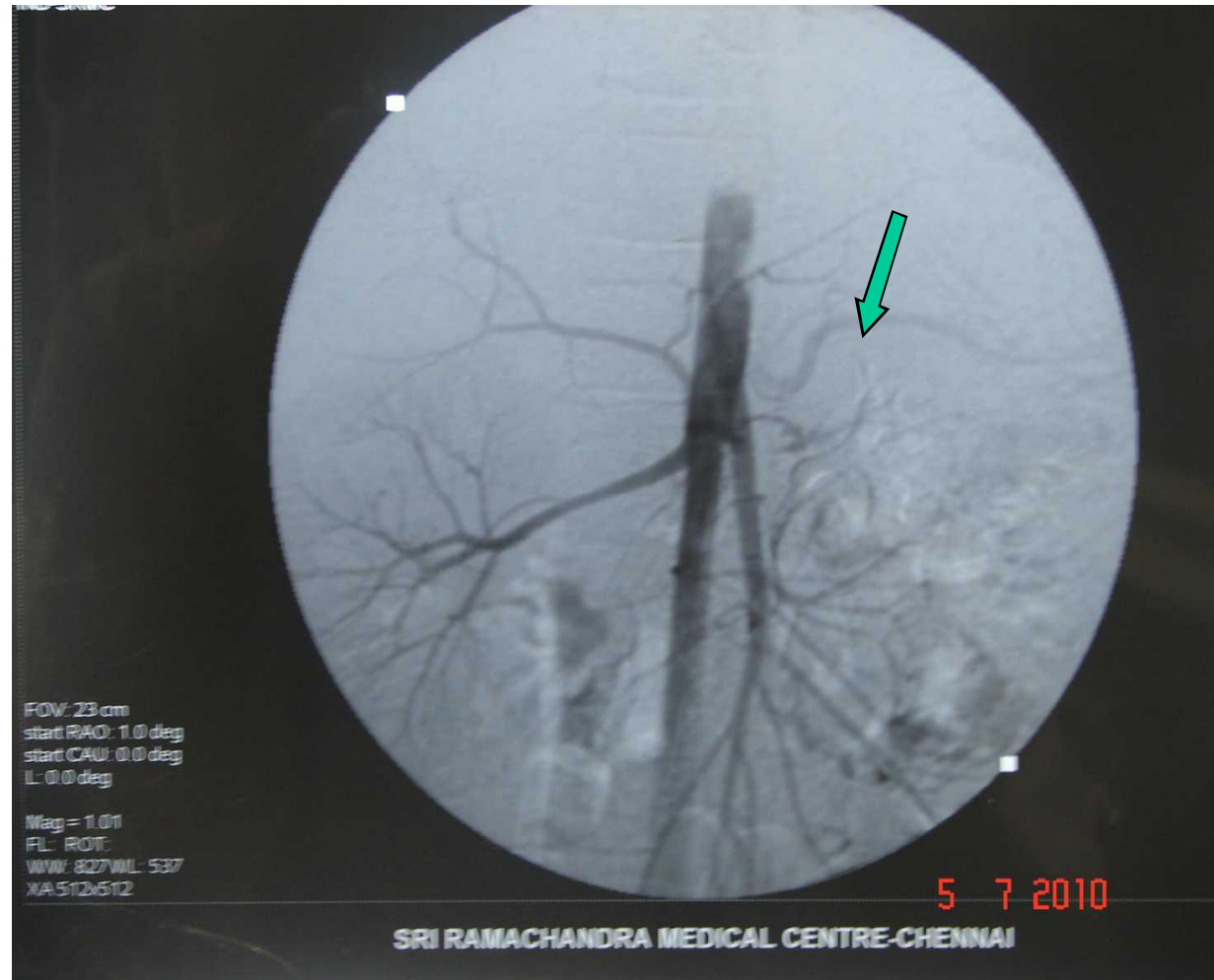
ANGIOGRAPHY REPORT

- Left MCA bifurcation aneurysm.
- Anterior Communicating Artery – Bleb
- Left renal artery occlusion with collaterals from pericapsular region

Cerebral Angiogram



Renal Angiogram

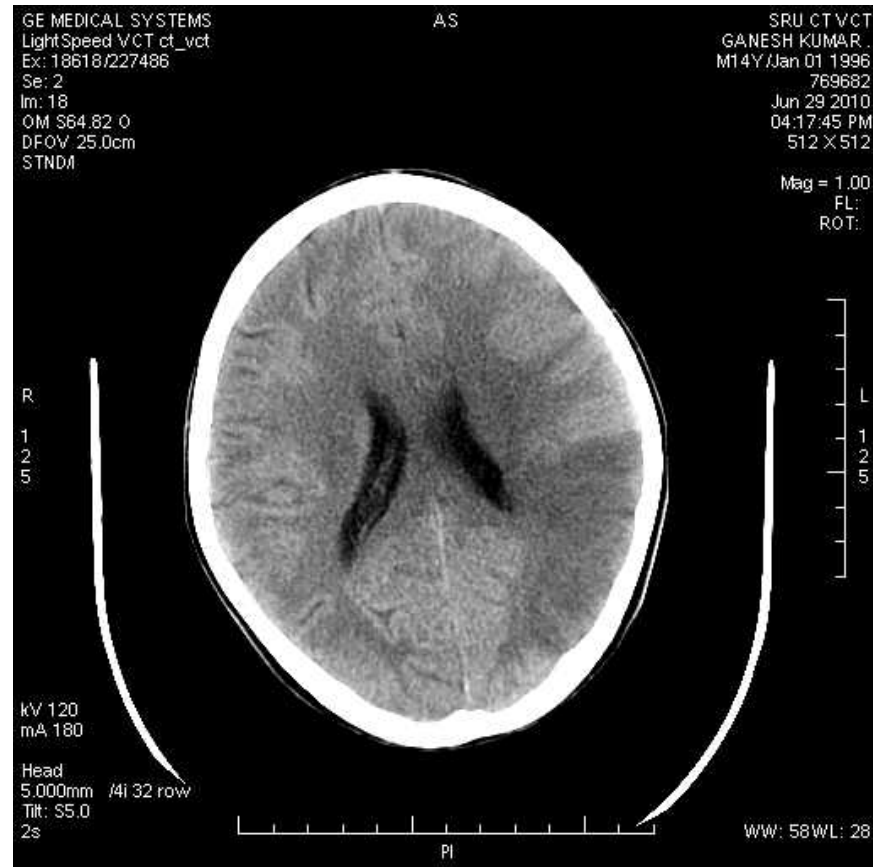




COURSE IN SRMC

- Developed speech disturbance which was typical of Nominal/ Conduction aphasia;
 - No other deficits;
 - CT scan Brain – Infarction of left infra temporal & parietal lobe
 - Possibly secondary to vasospasm
 - Speech Therapy commenced
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CT BRAIN



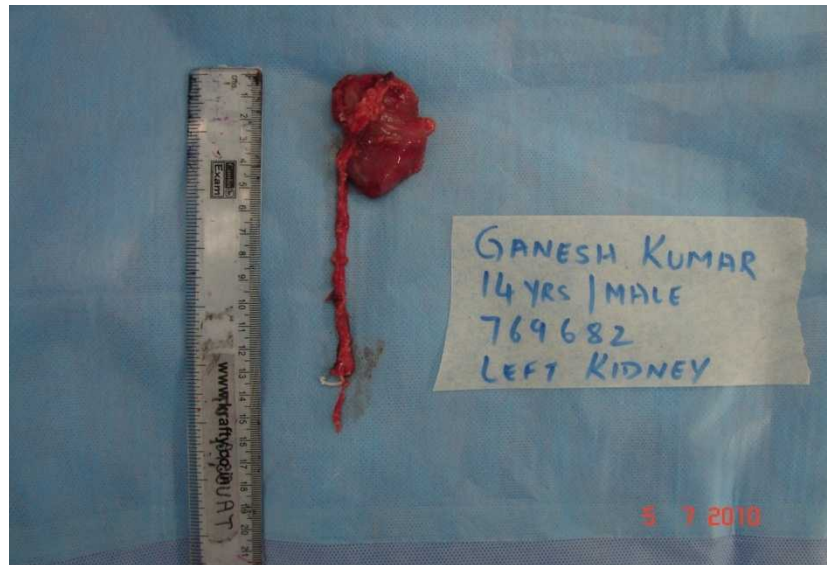


Further management

- After adequate hydration and Acetylcysteine peri-procedure, Endo-coiling of aneurysm was done
- Discharged on Nimodipine, Metoprolol, Alpha Methyldopa and Enalapril
- Possible diagnosis of fibromuscular dysplasia
Aneurysm ? Due to FMD ? Due to hypertension

Operative management

- Laparoscopic nephrectomy was done 1 month later





Histopathology

- Thickened intra lobar arteries.
- The renal arteries and its branches show hyperplasia of tunica media and narrowing of the lumina suggestive of fibromuscular dysplasia
- On follow up anti hypertensives were tapered and was on a single drug



Renovascular Hypertension

- RVH is hypertension resulting from a renal arterial lesion that is relieved by correction of the offending lesion or removal of the kidney

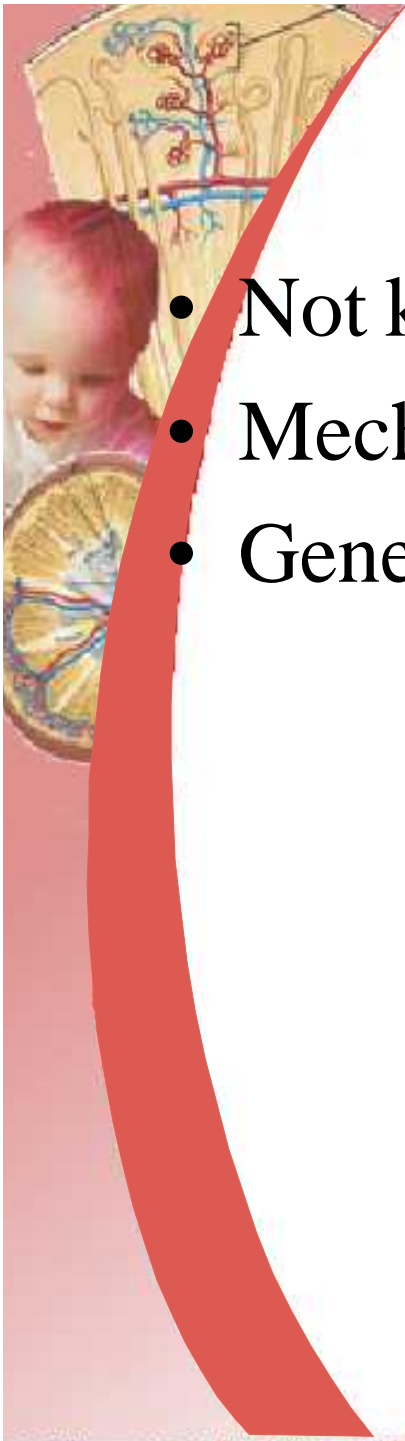


FIBROMUSCULAR DYSPLASIA (FMD)

- Formerly called as fibromuscular fibroplasia
- Group of idiopathic segmental non-atherosclerotic non-inflammatory disease of the musculature arterial walls leading to stenosis of small and medium sized arteries commonly involving renal and carotid arteries.
- May be familial
- The prevalence is about 4 in 1000 for symptomatic FMD in renal arteries and about half that for Carotoids.

AETIOLOGY

- Not known but may be environmental
- Mechanical mobility of kidneys
- Genetic factors



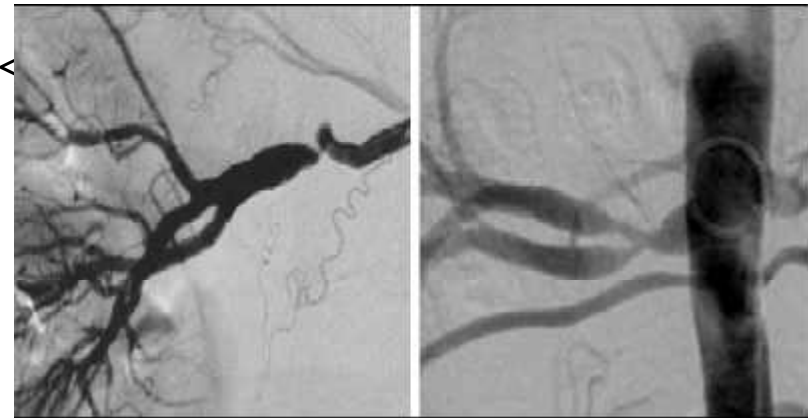


PATHOLOGICAL CLASSIFICATION

- Based on dominant arterial wall layer involved.
 1. Intimal (5%) – irregularly arranged mesenchymal cells within loose matrix of endothelial cells and fragmented internal elastic lamina
 2. Medial (85%)- homogenous collar of elastic tissue with multiple stenosis and aneurisms
 3. Perimedial (10%)- excessive tissue deposition at the junction of media and adventitia
- All types may coexist in the same patient
- Significant number of patients may not have a histopathology as they would have been asymptomatic or undergone only angioplasty.

ANGIOGRAPHIC CLASSIFICATION

- Multifocal with string of bead appearance seen mostly in women between 30 and 50 years involving distal and middle 2/3th of renal artery (62%)
- Tubular type (14%)
- Focal type with solitary < 1cm (7%)
- Mixed type (17%)





CLINICAL FEATURES

- Renal artery involvement (58%),
- Cervico-cranial arteries (32%) and others (10%)
- Macroaneurysms and dissections common
- Renal involvement presents with hypertension flank bruit, normal creatinine and rarely infarction
- Cervical headache, dizziness, brain ischemia,, embolus thrombosis, subarachnoid hemorrhage due aneurismal rupture
- Others- mesentric ischemia, multiorgan failure, limb claudication etc.



DIFFERENTIAL DIAGNOSIS

- Vascular Ehlers Danlos syndrome
- Williams syndrome
- Type 1 neurofibromatosis

Older individual

- Atherosclerosis
- Takayasu arteritis

DIAGNOSIS

- Ultrasound
- MRI angiography
- CT angiography
- **GOLD STANDARD** Intra arterial angiogram with digital subtraction justified if proceeding to revascularization



MANAGEMENT

- No hypertension observe
- Anti hypertensive drugs – Ace inhibitors, angiotensin receptor blockade, Calcium channel blockers, beta blockers
- Malignant hypertension – Revascularization – angioplasty and bailout stent
- Segmental arteries involved surgical reconstruction
- Carotid artery – anticoagulant, angioplasty or surgical repair
- Ruptured intracerebral aneurysm - - microvascular neurosurgical clipping and endovascular coiling





OUTCOME

- Multivessel disease may progress rapidly but long term outcome not known

REFERENCES

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TAKE HOME MESSAGE

- Childhood hypertension is usually secondary, and needs extensive evaluation
- It may be reversible when appropriately treated early and complications may be prevented