We at Max-Saket recently did two consecutive implantable ventricular assist devices for end stage heart failure in last three weeks. These are Delhi’s first and second devices.

Treatments available for end stage heart failure is either heart transplant or ventricular assist device. As the paucity of organ donation donor hearts are not available. In this situation when life expectancy is limited or patient cannot be weaned off from temporary mechanical support, the solution left is implantable LVADs. Both of these patients were young (late 30’s) known cases of dilated cardiomyopathy (DCM) for more than 2 years. One had AICD placed for more than one and a half year for LBBB. He was admitted to another hospital in acute on chronic heart failure. He went into multi organ failure and put on ventilator and inotropes. We transferred the patient to our hospital and optimized with medical therapy and temporary mechanical support. He could not be weaned off support and then Heart Mate II was implanted. Patient got better and was discharged home. Another young man was airlifted from another state for acute decomposition. Though he has a history of DCM for the last two years but because of narrow QRS complex AICD was not offered. He presented in low perfusion state with hepatic and renal dysfunction. Heart Mate II was implanted few days ago. His cardiac output improved and all organ functions got better.

This is a reliable and long term therapy which improves organ perfusion and thus quality of life. Life expectancy of such subset is very poor if not treated in time and they die in 6 months to one year. More than 28000 LVADs have been implanted globally and 10000 patients are on device at present. Out of these, more than 500 patients are on this device for more than 10 years.
Our comprehensive heart transplant services include medical therapy, FDA-regulated devices (LVAD), ECMO, Ambulatory Balloon pumps and heart transplantation. Our specialized team would use innovative technology and therapies to treat patients with heart failure.

Dr. Kewal Krishan, Program In-charge, Heart Transplant & Ventricular Assist Devices, Senior Consultant Cardiac Surgeon, Max Super Speciality Hospital, Saket, has done four years of advanced clinical fellowship at world’s top hospitals including Mayo Clinic, Rochester, USA and Mount Sinai Medical Centre, New York, USA where he gained expertise in advanced therapies. He was trained by internationally renowned surgeons for Heart Transplant and Ventricular Assist Devices. Dr. Kewal is one of a handful surgeon in India who is formally trained in all aspects of heart transplantation. He was trained intensively in the entire spectrum of ventricular assist devices including bridge to transplant, short term and long term devices and destination therapy. He has many publications in international journals to his name in this field including innovative techniques in ventricular assist devices. I am thankful to Dr. Rajneesh Malhotra for his continuous efforts helping me to build heart failure program. I sincerely thanks to Dr. KK Talwar and Dr. Viveka Kumar for all the guidance and patronage time to time. It’s Dr. Roopa Salwan’s efforts that guided and assured patient’s family of best possible care at our centre to save one’s life.

It’s an organized effort by heart failure experts to provide quality care for patients suffering from acute and chronic heart failure at Max Saket. Our mission is to enhance quality and duration of life in those with end stage heart failure.

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**Men who gave kidneys to wives, cheered!**

- 80% of living donors are women but they rarely get organs themselves: docs

Doctors at a city hospital felicitated husbands who had donated their kidney to their wives. They said, while 70-80% of living donors are women, only a few of them get a kidney should they, themselves, suffer from organ failure.

“Since September 2013, we have conducted 236 kidney transplants. Only 56 recipients were women. Most other women patients never came back after being diagnosed with kidney failure, either due to lack of donor or financial reasons,” said Dr Dinesh Khullar, director, nephrology and kidney transplant medicine, at Max Hospital, Saket. Of the 56 transplants, mothers (13) were the most common donors followed by husbands (12) and fathers (6). “We honoured husbands who donated kidneys to their wives to make them feel proud of their sacrifice and to inspire others,” Dr Khullar said.

TOI spoke to one such husband, Pawan Kumar Somani, who donated his kidney despite suffering from a debilitating disease. “How could I let her die? As a husband, it was my duty to do everything that I could. What’s more, doctors told me categorically that donating a kidney does not impact health,” Somani said. Ranvir Singh took voluntary retirement from UP police to arrange money for his wife’s transplant. He also donated the organ.

A study conducted by doctors at PGIMER, Chandigarh, shows donating a kidney to a spouse is associated with improved interpersonal relationships in the family. “Higher incidence of kidney disease in men, fear of losing the earning member and perception of renal donation as an extension of responsibility to family have been cited as reasons for female preponderance among living donors,” states the study, published in Indian Journal of Nephrology.

The Transplantation of Human Organs Act 1994 grants legal validity to the act of organ donation. “Before the law was passed, a lot of paperwork was required to prove that the woman was donating her organ of her free will,” said a doctor.

Risk factors for kidney damage are equally common among women. “There is need to root out the idea that donating a kidney can cause health problems among working men. Also, women should not be treated as second class citizens,” an expert said.

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Dr. Dinesh Khullar
Director & Head of Department
Nephrology & Kidney Transplant Medicine
Max Super Speciality Hospital, Saket
**Removal of intrahepatic bile duct stones by holmium laser lithotripsy** - A novel technique for a complex disease

**PRESENTATION**
A 52 years African national presented with 7 months history of right upper abdominal pain with nausea, vomiting and anorexia. She denied associated fever or jaundice. She underwent laparoscopic cholecystectomy and radical hysterectomy (for carcinoma cervix) 5 & 4 years ago, respectively. A recent abdominal ultrasound in her country was suggestive of hepatolithiasis.

On examination, she was thin built and there was no icterus. Abdominal examination also did not reveal abnormality except for scars of previous surgery. Her hemogram and kidney function tests were within normal range whereas the liver enzymes were elevated in a cholestatic pattern.

Abdominal MRI with MRCP to delineate the extent of hepatolithiasis and for treatment planning was suggestive of type I E (R+L) disease (Figure 1). There was an extensive involvement of both hepatic lobes. There was no associated liver atrophy. The diameter of the most peripheral dilated biliary ductules was reported to be 2-3 mm thereby making them suitable for access by a flexible ureterorenoscope (Karl Storz Flex 2). There are several other features that made this instrument ideal for this purpose - working length of 67 cm, external diameter 7.5 F, 270 degree deflection of distal tip and 1.7 F basket can be introduced.

A cholangioscopic (utilizing flexible ureterorenoscope) holmium laser (Lumenis 100 watt) lithotripsy with complete clearance of stones and hepaticejunostomy was planned.

**TECHNIQUE**
The peritoneal cavity was accessed through a right subcostal incision. The common bile duct was isolated and choledochotomy made between stay sutures. All accessible stones were removed in the standard manner using Desjardin’s forceps. Through the choledochotomy, a flexible ureteroscope (Karl Storz Flex 2, external diameter 7 F) was passed into the common duct and complete cholangioscopic examination performed. Then a 200 micron holmium laser fiber was introduced through the ureteroscope and all stones were pulverized. The larger stone fragments were removed with a basket (Ngage nitinol stone extractor, size 1.7 F; 115 cm length; Cook Medical) while the smaller stones were flushed out. Once the common duct was completely cleared of all stones, the right and the left biliary systems were systematically entered and all stones removed in a similar fashion from the subsegmental ducts and even beyond (Figure 2).

Finally the ureteroscope was passed antegrade into the duodenum to rule out any stone fragments in the distal common duct. The procedure was then completed by fashioning a choledochojenojunostomy (Roux loop of 60 cm length; single layer interrupted 4-0 PDS sutures) and a jejunojejunostomy.

Postoperative course was uncomplicated. The LFTs revealed a normalizing trend and the post operative imaging was not suggestive of any residual stone disease.

**DISCUSSION**
Hepatolithiasis is presence of gallstones in the bile ducts proximal to the confluence of right and left hepatic ducts. Patients may be asymptomatic, diagnosed incidentally on abdominal imaging or may present with pain right upper quadrant or epigastrium, jaundice and fever. Up to 10% of patients may have co-existing cholangiocarcinoma.

The goals for the treatment of hepatolithiasis are complete clearance of stones, resection of stricture bile duct & atrophic liver tissue and reconstruction of bile drainage.

The treatment of hepatolithiasis continues to be complex and difficult. The different accepted/ described management options for stone clearance include endoscopic retrograde cholangiography (ERC), percutaneous transhepatic cholangioscopic stone lithotripsy (PTCSL) and biliary enteric anastomosis. Liver resection is indicated in patients with isolated left hepatolithiasis, associated atrophic segments and/ or cholangiocarcinoma.

Our patient therefore needed a modality that would result in complete clearance of stones. Of the various techniques, ERC was deemed unsuitable in view of the extensive disease. The equipment/ experience with PTCSL is limited at most centres. Therefore based on our past experience of holmium laser lithotripsy for complex bile duct stones, we opted for this technique for the management of hepatolithiasis.

There are several advantages of this technique:
- Flexible ureterorenoscope with an external diameter of 7.5 F can access even the most peripheral ducts.
- Holmium laser as an energy sources results in complete pulverization of stones thereby facilitating an complete and expeditious clearance of stones.
- Complete clearance of the biliary ductal system with a single intervention.
- The entire biliary system can be visualized directly and lesions suspicious of concomitant cholangiocarcinoma can be biopsied.
- Equipment is available in all urology units in a tertiary care hospital. A separate choledochoscope with limited usage and versatility therefore is not required.

**Figure 1.** Stones in dilated intra and extra hepatic biliary ductal system of both lobes of liver **Figure 2.** Tip of ureteroscope seen through the liver parenchyma (arrow)
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Dr. Rajkumari B. Dhar (Visiting Consultant, Rheumatology)  Dr. Prerit Mathur (Programme Coordinator - Orthopaedics)
ALPSA v/s Perthe’s lesion (bankart variant) with slap 2 tear and GAGL – Max Hospitals’ first MR arthrographic study of Shoulder joint

- 41 year old male with history of traumatic dislocation of right shoulder 7 years ago.
- Since has had multiple spontaneous anterior subluxations within last 5 years, consistent with shoulder instability.

**MRI SHOULDER FINDINGS**
- Non arthrographic MRI demonstrates anterior labral tear with a fluid cleft along the undersurface of labrum and the glenoid rim suggesting a Perthes lesion which is a non-osseous Bankart variant.
- A doubtful SLAP tear is also seen.

**ARTHROGRAPHIC MRI**
- Arthrographic MRI clearly differentiates the thin flimsy diminutive labrum from the thickened rolled up middle glenohumeral ligament (MGHL), thickened anterior capsule and the large pouch of sleeve periosteum of anterior glenoid suggesting an ALPSA lesion (Anterior Labral Periosteal Sleeve Avulsion).
- Confirms a SLAP 2 tear.
- Shows avulsion of the glenoid attachment of inferior glenohumeral ligament (G AHL).

See more at: http://radsource.us/glenohumeral-instability/

Dr. Nafisa Shakir Batta, Dr. Monu Singh, Dr. Gaurav Mittal, Dr. Bharat Aggarwal
Department of Radiology & Orthopaedics
Max Super Speciality Hospital, Saket

**BANKART VARIANTS**

Perthe’s lesion
- First described by Georg Clemens Perthes
- Defined as detachment of the anterior labrum 3-6 o’clock, with medially stripped but intact periosteum

ALPSA lesion
- ALPSA = Anterior Labral Periosteal Sleeve Avulsion
- Medially displaced labroligamentous complex with absence of the labrum on the glenoid rim

See more at: http://radsource.us/glenohumeral-instability/
WHY DO DIRECT ARTHROGRAPHY OF SHOULDER?

- MR arthrography has emerged as an important technique in the assessment of selected musculoskeletal disorders.
- A plethora of studies have assessed this technique for the evaluation of shoulder, knee, wrist, elbow, ankle, and hip pathology.
- Traditionally used flouroscopic guidance for shoulder arthrogram has been replaced by ultrasound guided joint injection, which is far more accurate, has less chances of periarticular leak and being posterior approach gives a clean diagnostic field.
- Direct MR arthrography utilizing dilute gadolinium contrast agent, has many advantages: consistent joint distention with contrast medium; markedly improved delineation of key intra-articular structures and consequently improved detection of surgically correctable pathology; more invasive and expensive procedures such as diagnostic arthroscopy maybe obviated.
- It gives excellent resolution of rotator cuff tears, labral pathologies, ligament and capsular injuries.

Findings were confirmed intraop, and coracoid process harvestd with attachment of conjoint tendon as graft. Graft with passed through subscapularis ad fixed with screws to anteroinferior glenoid labrum.

Eat right.
Eat healthy

Ritika Samaddar stocks a variety of healthy fare at home: nuts, sprouts, fruits, soya milk, lassi.

All the meals in my home are planned keeping the children in mind. To ensure my family gets nutrition from all food groups, we include dairy, eggs, fruit and a teaspoon of flaxseeds every day. There are several new kinds of diets that people follow today, however a restrictive diet may be necessary if you’re lactose intolerant or diabetic or have any short term goals, but insufficient to meet nutritional goals.

WHAT’S ON MY PLATE

I start my day with ¼ tsp cinammon in boiled water, with ¼ tsp honey and a handful of nuts. An hour of aerobics or yoga or zumba later, I have a cup of tea. I never have coffee/tea with my meals as it leaches iron and calcium from the bones. Twice a week, I fast and only eat fruits and a glass of milk. Other days, being a dairy lover, I have milk and an egg-white omelette for breakfast. Lunch is mostly salads with sprouts, brown rice, or pasta and chicken. Also, I have lassi or a cup of curd afterwards. I’m prone to irritable bowel syndrome, so curd soothes the stomach. I’m a Bengali and we love fish and its on my plate 4 times a week. Else, it’s chicken or mutton. I have no carbs (no roti or rice or grains) at night, only vegetables or fresh fruit and veggie salad with fish or meat. I always keep a stock of soya nuts, roasted gram, nuts and a box of seasonal fruits to snack on and a flask of nimbu pani. Small, frequent meals work for me. Back home, at 6.30 pm, we have soups in winter, or fresh mixed veg/fruit juice in summer.

HOW I EAT A RAINBOW

Through seasonal fruits, that’s easier than preparing salads. I grow spinach, chillies, coriander and plan to plant arsley, thyme and other herbs too.

MY KITCHEN IS ALWAYS STOCKED WITH

Wholesome snacks. With a daughter taking her Board exams, I ensure a variety of healthy fare at home—sprouts, mixed nuts (walnuts, dates, prunes, raisins) and small packs of fruit juices, soya milk and lassi.

SECRET INDULGENCE

Indian sweets. I love misthi doi, sandesh and rasagulla and have it once or twice a week.

Ms. Ritika Samaddar
Regional Head, Dietetics
Max Healthcare, New Delhi

QUICK TIP

Add yoghurt to fish curry, matar paneer or mushroom dishes. Yoghurt is healthy; it also makes the gravy thick and gives the dish a unique flavour.

Source: Prevention, April 2015
Need for awareness on rare disorder

Bindu Shajan Perappadan

NEW DELHI: A year ago, two stunted brothers afflicted with Hunter syndrome were rushed to a hospital in South Delhi.

Puneet Agarwal, the head of neurology unit and the principal consultant at Max Super Speciality Hospital in Saket, reviewed their condition and discovered that one of them — 22-year-old Raja — was suffering from fits and seizures due to accumulation of hydrocephalous fluid in the brain and the thinning of brain nerves.

"In normal adults, the fluid is distributed from the brain. However in Raja’s case, since brain growth had stopped, the fluid had gathered inside the brain, causing seizures. A similar issue had affected Prince [his brother] five years back. Prince [24] was also managed by us," Dr. Agarwal added. Both siblings have now been put on oral medication for life and are doing well, he said.

It is not just the brain in the case of these brothers, but other organs and their body too that refused to grow after a while.

Brothers Raja (22) and Prince (24) suffer from Mucopolysaccharidosis type II or Hunter Syndrome

“This is due to a rare genetic disorder called Mucopolysaccharidosis type II [MPS II], commonly known as Hunter Syndrome. We want to highlight this case because awareness about the syndrome is very low and we hope that talking about the brothers and discussing this health condition will ensure that those affected by the same are brought to the hospital and adequate medical support is provided to them at the earliest," said Dr. Agarwal.

MPS II is a lysosomal storage disorder caused by mutation of a gene resulting in deficiency of the lysosomal enzyme in the body. As a result, the body doesn’t grow normally. While symptoms are different in each patient, some common ones include severe airway obstruction, skeletal deformities, cardiomyopathy and neurological decline.

The disorder slows the physical and mental growth of the patients too. This is why Prince and Raja have a mental age of a 10-year-old child. Their physical growth has also been sluggish.

Their father Manjit Singh has started a society called Lysosomal Storage Disorders Support Society (LSDSS). Started in 2010, the society has registered 592 children with lysosomal disorders, of which 230 suffer from MPS II.
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Single stage repair of a rarely encountered complex congenital heart disease with multilevel left side obstruction - A case of Shone's anomaly

INTRODUCTION

Complex congenital heart disease with multilevel obstruction on left side is relatively rare and challenging disease to manage. Shone's syndrome is complex congenital heart disease which presents as obstruction at multiple level on left side of heart. Four potentially obstructive lesions of Shone's complex as originally described by Shone et al constitute: supramitral ring, parachute deformity of the mitral valve, subaortic stenosis and coarctation of aorta (Figure 1). These anomalies tend to coexist, but the severity and predominance of each individual lesion may vary. In the original article of Shone et al only 2 patients exhibited all four aspects of Shone's complex. The remaining can be regarded as formes frustes of Shone's anomaly usually with absence of supramitral ring. In clinical practice the definition of Shone's anomaly has been extended beyond the original Shone's complex, to encompass patients with additional forms of left heart anomalies, such as mitral and aortic valvular lesions and supraaortic stenosis. Varying degrees of endocardial fibroelastosis and hypoplasia of the mitral valve and left ventricle (LV) cavity add to the management challenge.

CASE REPORT

We present here a case of Shone’s anomaly with multilevel obstruction on left side. This 3 month child presented to us with history of failure to thrive and congestive heart failure (CHF). On examination pulses were feeble in both lower limbs while upper limb pulse was well felt with pulse rate of 140/min. The blood pressure was 98/50 mmHg in right upper limb. The first heart sound was loud with a mid diastolic murmur of grade III at apex and ejection systolic murmur of grade III prominent at second and third intercostals spaces radiating to carotid. ECG showed biventricular hypertrophy and CXR showed CT ratio of 0.65. Two dimensional echocardiogram showed severe mitral stenosis at valvular and subvalvular level with a mean gradient of 11 mmHg in right upper limb. The first heart sound was loud with a mid diastolic murmur of grade III at apex and ejection systolic murmur of grade III prominent at second and third intercostals spaces radiating to carotid. ECG showed biventricular hypertrophy and CXR showed CT ratio of 0.65. Two dimensional echocardiogram showed severe mitral stenosis at valvular and subvalvular level with a mean gradient of 11 mmHg, subaortic membrane and accessory tricuspid valve tissue causing left ventricular outflow tract (LVOT) obstruction, dynamic midcavity left ventricle (LV) obstruction, moderate size ventricle septal defect (VSD) (Figure 2) and severe pulmonary artery hypertension (PAH) and coarctation of aorta (CoA) with moderate size patent ductus arteriosus (PDA) (Figure 3). Following a failed attempt at balloon dilatation of the coarctation the child was planned for a single stage surgical correction of left side inflow and outflow obstruction and VSD closure. Intraoperative invasive arterial pressure monitoring of both upper and lower limb showed difference of >35 mm Hg across the coarctation.

After sternotomy ascending aorta, aortic arch along with branches and descending thoracic aorta was mobilised extensively. She was then placed on cardiopulmonary bypass with moderate hypothermia. Narrowed segment of aorta was excised and extended end to end anastomosis of arch with descending thoracic aorta (DTA) done. After that cross-clamp was reapplied proximally on the aorta and heart arrested with cold blood cardioplegia. Right atrium opened and intracardiac repair accomplished. Accessory tricuspid valve tissue and thick subaortic membrane excised in toto. LVOT myotomy was done anterosuperiorly to enlarge LVOT in view of dynamic midcavity obstruction. VSD was closed with Dacron patch. Interaltrial septum was opened and mitral valve exposed and thick supramitral ridge excised. Mitral valve repaired with thinning of PML and splitting of tissue between chordae and both papillary muscle heads. After satisfactory repair intertrial septum and right atrium closed and cross clamp removed. Intraoperative echocardiogram after coming off bypass showed mean gradient of 8 mmHg across repaired segment of CoA and laminar flow in mitral valve with trivial MR/MS and no residual VSD and LVOT. She was electively ventilated for 48 hr in ICU and then extubated on postoperative day 2. She is doing fine and pre-discharge echo showed well opened arch of aorta with minimal gradient across repaired segment of CoA (Figure 4) and mitral valve (4.5 mmHg).

Dr. Praveer Sinha, Dr. K.S. Dagar
Dr. Lokesh Jaiswal, Dr. Shoumik Pal
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DISCUSSION

Shone’s complex was initially described by Shone ET AL in 1963. As discussed above it consists of four major components originally presented by Shone ET AL. Supramitral ring is a circumferential ridge or membrane which arises from left atrial wall overlying the mitral valve and is frequently attached to the mitral valve. The ring may range from a thin membrane to a thick discrete fibrous ridge. There is various degree of mitral inflow obstruction which may be due to restricted opening in membrane, encroachment of ring onto mid anterior mitral leaflet which severely restrict leaflet excursion and its opening or ring may be large and protrude into the mitral valve inflow causing obstruction. Parachute mitral valve is defined as a unifocal attachment of mitral valve chordae independent of the number of papillary muscles. A true parachute mitral valve is characterised by attachment of the chordae to a single or fused papillary muscles. The unifocal attachment of the chordae results in restricted valve opening and subvalvular obstruction. LVOTO in Shone’s complex is mostly due to discrete subaortic membrane with varying degree of obstruction. However there can be long tunnel like severe subaortic obstruction or obstructive bicuspid valve.

For complex congenital heart disease with multilevel left sided obstruction like Shone’s syndrome usually multiple stage treatment is needed with operative mortality ranging from 16 to 24% with variable late mortality and morbidity. The management strategy of this complex population is hampered by the variable mode of presentation and differing severity of individual lesions. Data from previous studies have shown that outcome in this population is related to predominance and severity of mitral component of the disease, associated pulmonary hypertension and need for multiple surgical intervention. Severity and type of mitral valve disease mainly affect the long term outcome of treatment.

Recent studies have shown favourable late outcome and reduced operative mortality affected mainly by the severity of mitral valve disease and degree of left ventricular hypoplasia and need for multiple operative procedure. The improvement in outcomes in recent studies is mainly due to adoption of reparative strategy for mitral valve along with routine intraoperative transesophageal echocardiography and replacement only in case of failed repair.

Single stage repair of complex congenital heart disease is a challenge in itself. Most of the patients present during the neonatal period with a critical coarctation of the aorta and minimal signs of left ventricular inflow pathology. This is of importance since the predominance of outflow tract symptoms may mask other potentially critical inflow
lesions. Therefore, it is critical that an aggressive diagnostic evaluation be undertaken for patients that present with a coarctation in the setting of even trivial mitral valve pathology. In our case we planned for balloon dilatation of CoA followed by VSD closure, MV repair and release of LVOTO. As coarctation could not be relieved successfully with balloon dilatation, we had two options at this stage – is repair of coarctation via thoracotomy followed by intracardiac repair at second stage and repair of both CoA and relief of significant multiple level obstruction in same stage. As studies have shown multiple operation and significant mitral valve disease to be independent risk factor for long term outcome and operative mortality, we planned for single stage repair of this complex congenital heart disease. For infants that are diagnosed prenatally with Shone’s syndrome, it is recommended that delivery take place in a tertiary care hospital with transfer to the Neonatal Intensive Care Unit as soon as possible to initiate cardiology evaluation and medical interventions. When coarctation is present, prostaglandin E infusion is started as soon as possible to keep the ductus arteriosus open prior to surgery. Intervention shortly after birth to repair coarctation of the aorta is usually necessary. Multiple surgical interventions may be needed to relieve left-sided obstructions.

In conclusion, operative management of patients diagnosed with Shone complex has improved with a clearer understanding of the valvular pathology and expertise with reparative techniques. Significant obstructive lesions of both left inflow and outflow tract should be addressed initially with reparative techniques whenever possible with replacement being reserved for failure of repair. Single stage repair though challenging has the potential to offer good early and long term results in this complex and demanding subset.

REFERENCES

"I must be eating right. I'm narrow at the top and wide at the bottom, just like the Food Pyramid!"

"If I eat one piece of cake instead of two pieces of pie, I can save 800 calories. Finally, a diet I can live with!"

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