Letter to the Editor

Adverse Outcome of Coarctation Stenting in Patients With Turner Syndrome

To the Editor

We read with interest the paper by van den Hoven et al. regarding adverse outcomes of coarctation stenting in patients with Turner syndrome [1]. There is no doubt that this specific subset of patients presents a different risk profile in contrast to the usual coarctation population. Due to the fragile aortic wall, a higher than normal complication rate is to be expected if "standard techniques" are used. Standard protocols in a usual coarctation population yield a complication rate of 0-7%, depending on the degree of gradient relief that is pursued. The adequate use of covered stents can nearly abolish the risk for blood extravasation but only within the area of aortic wall covered by the stent. However, many centers use covered stents as if it were simply an extension of a bare metal stent. In order to get all the advantages of a covered stent among which an added safety margin, adapted techniques are required [2]. If a tear or rupture of the aortic wall occurs where the stent makes contact with the wall, the covering at that area should prevent blood extravasation; this is the minimum one expects from a covered stent. However, a tear may stretch beyond this initial contact zone making additional safety precautions necessary. This can be accomplished by sealing off the surrounding areas where a potential leak can occur; here the operator should aim for good apposition of the distal and proximal ends of the stent against the walls of the upstream and downstream aorta. This typically requires flaring of the edges of the covered stent with bigger balloons. Such sealing requires a longer and wider stent than typically used for bare metal stenting; a covered stent ideally should bridge from healthy tissue to healthy tissue with the pathological part well covered in-between. The areas of post stenotic dilation may reach diameters of 25mm or more! For some of these sizes it may be wise to use custom made or self-expandable stents. Frequently, state of the art stenting may require initial balloon interrogation, a balloon to deliver the stent, additional (low pressure) balloons to flair at the upstream and downstream ends, and additional high

pressure balloons to post-dilate the stenotic hypoplastic segment. Ideally the edges of the stent should not be pushed into the wall, but gently apposed onto the wall to get adequate sealing without causing a local tear. Stents are not all alike: the metal edges of a stent should be atraumatic: a laser cut strut will have a thinner profile but sharper edges than a thicker rounded strut. The edges of a sharp strut may perforate the tissue causing a tear at the edge of a stent, leading to an aneurysm or a downstream dissection. The struts should be strong enough to avoid infolding as has been reported with some covered stents [3,4]. Delayed dilation several weeks after initial stent implantation may be the safer option in some patients: it allows some adaptation of the stretched wall thereby limiting the eventual tear, better sealing of the tissues surrounding the site of the expected vessel tear, and allows "spontaneous" thrombosis of collateral arteries. Such collaterals may bleed extensively into the mediastinal and pleural spaces if still patent when the narrowed aortic wall is torn; a covered stent will only protect the patient from bleeding from the aorta lumen. Bleeding from collateral arteries must be avoided: it may be self-limiting, but when profuse cannot be managed by percutaneous techniques and surgical rescue will be very difficult.

If significant bleeding from the aorta occurs, surgery most likely will be too late leading to enormous morbidity and mortality. It is probably faster and safer to solve the problem percutaneously by flaring the stent to seal, or by adding another longer and bigger stent to obtain additional sealing.

Techniques for redilation should also be adapted since the aortic wall may be avulsed off the edges of the stent causing an early or late severe dissection [5]. This can be avoided by using a balloon no longer than the stent and not too large. If any doubt about a tearing

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at the edges exists, it would be sensible to use a new covered stent over that edge to seal any possible leak or tear.

The post procedure coagulation protocol is still debated. However, as is illustrated by this series, the most serious complications are bleeding, dissection, and aneurysm formation, all of which are theoretically enhanced by anti-platelet drugs. The risks of such drugs appear to outweigh the presumed but unproven advantages.

The fact that local protocols and techniques were not specified in the report is disconcerting in view of the abovementioned and the serious nature of the complications experienced. Frequently standard techniques are based on bare stents, while correct usage of covered stents imply usage of longer, bigger stents with rounded edges, deployed by multiple balloons (interrogate, deliver, seal, dilate) in sometimes multiple sessions. From the data it is not clear how the stent lengths and diameters were selected. It is conceivable that stents were of adequate length to cover a stenosis, but not to seal a possible tear. The usage of covered stents has no effect against dissection at the edges, nor bleeding from collaterals.

In the report of van den Hoven no serious events occurred during the procedure, however two dissections occurred within hours-days after the intervention, which should remind us that careful monitoring in these patients is indicated. All complications are known and technically avoidable provided adapted techniques are used. The two late deaths probably reflect the natural history of the disease.

We agree with the authors that when discussing coarctation treatment with a Turner patient, the clinician should consider—as always—all reasonable options. Future studies will determine whether current medication, stateof-the-art surgery, or adapted percutaneous interventions will be the better option. Bjorn Cools, MD Pediatric and Congenital Cardiology UZ Leuven Leuven, Belgium

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